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LOBAR OBSTRUCTIVE EMPHYSEMA IN INFANCY TREATED BY LOBECTOMY

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LOCALIZED emphysema is commonplace in the presence of pulmonary infection[1-5] or an intrabronchial foreign body,[6, 7] but rarely has it been described in otherwise apparently normal infants. Lobar obstructive emphysema in infancy produced by an abnormality of the bronchus is being recognized with increasing frequency. The recent publication of several reports of successful lobectomy for obstructive emphysema in infants has made the recognition and treatment of this condition of greater interest to surgeons.

In 1939 Overstreet[8] reported the case of an infant with obstructive emphysema of the left upper lobe who was found to have deficient cartilage rings in the left upper lobe bronchus on post-mortem examination. Overstreet mentioned that a lobectomy had been performed in another hospital for a similar lesion in an infant without relief of symptoms. Other scattered descriptions of obstructive emphysema in infancy resulting from a bronchial abnormality appeared but on none of the patients was an operation performed.[9, 10]

Gross and Lewis,[11] in 1945, presented a case involving an infant with obstructive emphysema of the right upper lobe resulting from the absence of cartilage rings in the right upper lobe bronchus. This was associated with a pleural defect of the anterior mediastinum. Lobectomy was performed and the mediastinal defect repaired. Lewis and Potts[12] later added a similar case of an infant with an anterior mediastinal defect associated with emphysema of the right middle lobe. No bronchial abnormality was found. Lobectomy was performed but the mediastinal defect was not repaired. Leahy and Butsch[13] also were able to report a successful lobectomy for emphysema of the left upper lobe in an infant.

The first presentation of a series of infants with lobar obstructive emphysema believed to be on a congenital basis appeared in 1951 when Robertson and

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James[14] reported the cases of five infants with this condition. Three of the infants underwent lobectomies. Robertson and James believed that the obstructive emphysema present in these infants was due to a check valve mechanism in the bronchus of the involved lobe. In two of the patients the bronchial obstruction was thought to be produced by weakness of the bronchial wall or a flap of bronchial mucosa. However, in another infant pressure on the bronchus from an abnormal, dilated vein was felt to contribute to the obstruction. In two of the infants no abnormality of the bronchus was found.

Shaw[15] stated that abnormal flaccidity of the bronchus was responsible for the bronchial obstruction present in the two patients with obstructive emphysema upon whom he operated. In one of the patients a single segment was involved. In the other the entire lobe was emphysematous. Williams[16] has reported the case of an infant with emphysema of the right middle lobe. Lobectomy was performed but no bronchial abnormality was described.

Finally, Fischer, Potts and Holinger,[17] in 1952, presented five additional cases of infants with lobar obstructive emphysema for which lobectomy had been done. In two of the infants, in whom the left upper lobe was involved, the bronchial obstruction was believed to be related to pressure on the left bronchial tree by the ductus arteriosus. In one infant there was hypoplasia of the bronchial cartilages of the involved lobe. No definite bronchial obstruction was found in the other two infants. These authors first pointed out that in none of the cases reported by them or by other authors had a lower lobe been involved. They speculated that greater expiratory forces acting on the lower lobes prevented the trapping of air in them.

In the majority of the patients respiratory distress was noted soon after birth. The infants had wheezing respirations, dyspnea, and cyanosis. The onset of illness was rarely associated with definite evidence of respiratory infection. The infants did not improve materially with any therapy. They usually had increasing respiratory distress. It was necessary to operate upon some of the infants as emergencies. Lobectomy in these patients was lifesaving. All of the patients operated upon survived the operation and all were greatly improved with one exception. None of the infants was reported to have had further respiratory difficulty following operation with the exception of the patient mentioned by Overstreet. The diagnosis of obstructive emphysema unrelated to infection or to an intrabronchial foreign body was not difficult once the clinical and roentgenographic picture had been recognized. However, this form of obstructive emphysema was confused frequently with cystic disease of the involved lobe, pneumonia, or atelectasis of adjacent lobes.

The first infant in whom lobar obstructive emphysema was recognized in the University Hospital was admitted in May, 1950. Within a space of two years three additional infants with this condition were admitted. These infants presented the same clinical and roentgenographic features described by previous authors. In contrast to the experience of others, they continued to have respiratory symptoms following discharge from the hospital, although they were improved by lobectomy.

CASE REPORTS

CASE 1.—

History.—R. B., U. H. No. 682439, a 7-month-old white male infant, was admitted to the Pediatric Service of the University Hospital on May 23, 1950, with difficulty in breathing. The infant was born Nov. 7, 1949, following an uncomplicated pregnancy of eight months. He weighed 6 pounds, 6 ounces at birth. The infant breathed and cried spontaneously.

Soon after birth it was noted that he had some difficulty in breathing. Oxygen was not required and the infant nursed well. When he was three weeks old he developed wheezing and a dry cough which was aggravated by crying. The cough bore no relation to meals and occurred both day and night. Fever was not noted by the parents. The administration of penicillin and a sulfonamide made little change in the condition. The symptoms subsided somewhat from the seventh to the tenth week of age but recurred again with increasing severity. The administration of penicillin on several other occasions produced no change in the symptoms.

The infant continued to do poorly. Despite a good appetite, he gained weight slowly. Cyanosis was noted when he cried.

Family History.—Two older brothers, three years and sixteen months of age, were normal. There was no family history of congenital abnormalities or of allergy.

Physical Examination.—Temperature was 101° F. (rectally); blood pressure, 104/45; pulse, 130; respirations, 28; weight, 11 pounds, 13 ounces.

On examination the infant was inactive and underdeveloped. He cried weakly and was cyanotic when he cried. Respirations were slightly labored and expirations were prolonged. There was retraction of the thoracic cage during inspiration. The anterior-posterior diameter of the chest was increased. The right hemithorax was more prominent than the left. Breath sounds were decreased over the right hemithorax. A few coarse râles were heard at the right apex. The mediastinum was shifted to the left. The heart was normal. No other abnormalities were found.

Laboratory Data.—Kahn test was negative; hemoglobin, 11.2 Gm.; red blood cells, 4.64 million; white blood cells, 13,750 with 39 per cent polymorphonuclear leukocytes, 47 per cent lymphocytes, and 12 per cent monocytes; urine, normal findings; tuberculin, negative 1/1000; normal trypsin activity.

Roentgenograms.—Seven films of the chest exposed at another hospital from Dec. 5, 1949, through April 18, 1950, were all interpreted as showing emphysema of the right mid-lung field with a well-defined triangular area of increased density at the right base and patchy density in the right upper lung field. The left lung was normal. The mediastinum was shifted to the left. Chest films taken at the time of admission showed similar changes (Fig. 1). The infant was fluoroscoped. There was no mediastinal swing with respiration and no change in heart size.

Course.—It was felt that the infant had pneumonitis or atelectasis of the right upper lobe and the right lower lobe, although there was no fever. On one occasion an audible expiratory wheeze was noted. Excellent temporary relief was obtained with Adrenalin. The patient continued to have periodic respiratory distress which increased in frequency and duration. X-ray films of the chest demonstrated increase in the emphysema of the right mid-lung field and greater shift of the mediastinum to the left. It was at last realized that the infant's problem was one of obstructive emphysema rather than atelectasis or pneumonitis.

Bronchoscopy.—On June 19, 1950, he was anesthetized with ether and the bronchial tree examined through the 3.5 mm. bronchoscope. There was some edema and hyperemia of the entire bronchial tree. The bronchial walls approximated during expiration. No foreign body was found. No localized obstruction of the bronchial tree was noted.

Operation.—On June 29, 1950, a right thoracotomy was performed through the fourth intercostal space. Ether-oxygen anesthesia was administered through an endotracheal tube. When the chest was opened a tense, distended mass of lung presented. This filled the whole right hemithorax. It could not be compressed. The mass was delivered into the wound. The infant's somewhat precarious condition immediately improved. The compressed right upper lobe and right lower lobe now could be inflated easily by the anesthetist. There were no enlarged lymph nodes about the right middle lobe bronchus. No bronchial abnormality was noted. A right middle lobectomy was carried out easily by the individual ligation technique. The expanded right upper lobe and right lower lobe filled the chest. Anterior and posterior drainage tubes were inserted. The chest wall was closed in layers with silk. Blood loss was estimated to be 142 c.c. The infant was given 200 c.c. of whole blood during the operation. He was returned to the ward in excellent condition.

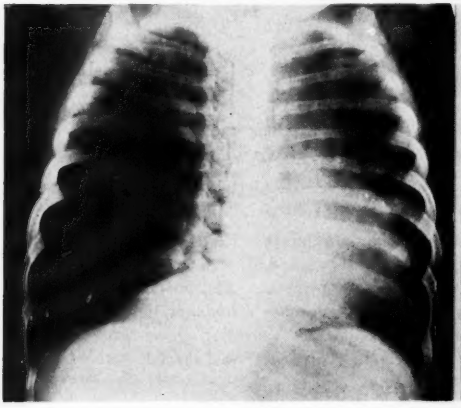

Fig. 1.

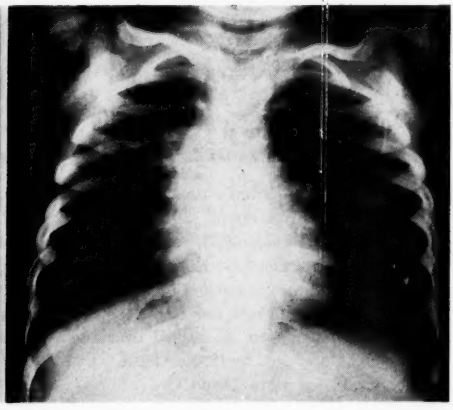

Fig. 2.

Fig. 1 (Case 1).—Preoperation roentgenogram exposed May 24, 1950. There is marked emphysema of the right middle lobe with compression of the right upper lobe and right lower lobe. The mediastinum is shifted to the left, and the right hemidiaphragm depressed.

Fig. 2 (Case 1).—Postoperation roentgenogram exposed Jan. 9, 1953. There is no emphysema. A small residual area of pleural reaction persists in the right paratracheal region.

Examination of the Specimen.—On gross examination there was uniform emphysema of the entire middle lobe (Fig. 3). The bronchus, although small in comparison with the greatly distended lobe, was not abnormal. Cartilage rings were present in the bronchial wall. On microscopic examination there was marked emphysema of the pulmonary parenchyma and dilatation of the smaller bronchi (Fig. 4). Slight, chronic bronchitis was present in isolated bronchi. No abnormality of the middle lobe bronchus was found in sections taken through it. Elastic tissue stains showed normal amounts of elastic tissue to be present.

Postoperative Course.—The infant's symptoms were completely relieved following operation. He became much more bright and active. The right upper lobe and right lower lobe expanded well and filled the right hemithorax, although the mediastinum was shifted slightly to the right. He was discharged on July 10, 1950. At this time respirations were normal.

Course Following Discharge.—The infant remained improved and developed in a normal manner. However, he had continued respiratory difficulties. On three occasions admission was necessary for dyspnea and wheezing respirations during an apparent acute respiratory infection. These symptoms responded to antibiotic therapy. In addition, the infant had frequent, milder episodes during which similar symptoms were present.

Bronchoscopy was repeated on Oct. 25, 1950. Again it was noted that the bronchi collapsed during expiration.

Fig. 3 (Case 1).—Photograph of the cut surface of the right middle lobe. There is marked emphysema of the entire lobe.

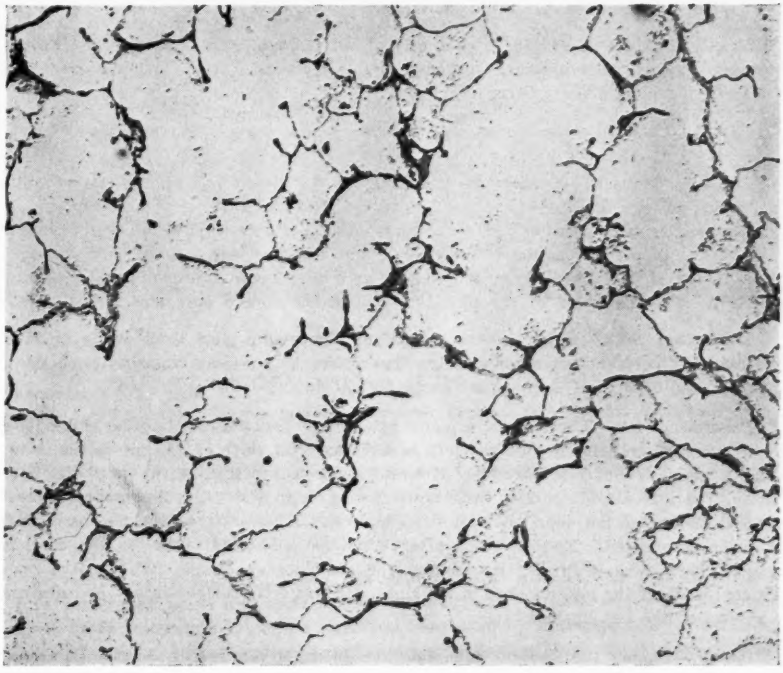

Fig. 4. (Case 1).—Photomicrograph of a section of the right middle lobe. Marked emphysema is present. There is no evidence of infection (×75).

X-ray views of the chest showed a triangular shadow of increased density in the right paratracheal area. This cleared gradually and was thought to represent reaction to operation rather than atelectasis.

The patient was last admitted Feb. 1, 1953. Admission was necessary for wheezing, coughing and difficult respiration. Adrenalin on one occasion gave temporary relief. The child quickly responded to penicillin and sulfadiazine. Because the symptoms resembled asthma, sensitivity studies were carried out but no specific protein sensitivity was found.

The infant was fluoroscoped and chest films were obtained Feb. 3, 1953. The diaphragms moved well. There appeared to be slight emphysema of the lung fields bilaterally but no unusual flattening of the diaphragms. There was no mediastinal shift with respiration. No emphysema had been seen in chest films obtained Jan. 9, 1953 (Fig. 2), and the findings at the time of admission may have been associated with the acute episode described.

CASE 2.—

History.—A. C., U. H. No. 712813, a white male infant 6 days old, was admitted to the Pediatric Service of the University Hospital Sept. 14, 1951, with difficulty in breathing and cyanosis. The mother's pregnancy was normal and the infant was delivered after an uncomplicated five-hour labor on Sept. 8, 1951. The infant weighed 6 pounds, 13 ounces. He breathed spontaneously and seemed normal.

During the first two post-partum days the infant had no respiratory difficulty. On the third day, when the mother's milk started to increase in amount, the infant choked on a feeding. On the fifth post-partum day the infant was returned to the nursery afer a feeding. He then became cyanotic and appeared to have obstructed respiration. The infant coughed up thick material which had the appearance of ingested milk and the condition improved. However, he continued to have rapid, labored respirations.

Family History.—A sibling, 3 years of age, was normal. The father gave a history of seasonal allergic episodes in which he had moderately severe asthma. There was no family history of congenital abnormalities.

Physical Examination.—Temperature was 99.4° F. (rectally); pulse, 140; respirations, 80 to 100.

The infant was active but was cyanotic. He was breathing rapidly and vigorously. Examination of the thorax showed active diaphragmatic and intercostal movements. The accessory muscles of respiration were being employed. Breath sounds were greatly diminished and resonance to percussion was increased over the left hemithorax. The mediastinum was shifted to the right. There were a few scattered râles heard over the right upper chest posteriorly. The remainder of the physical examination showed only normal findings.

Laboratory Data.—Kahn test was negative; hemoglobin, 15.8 Gm.; white blood cells, 22,500 with 44 per cent polymorphonuclear leukocytes, 46 per cent lymphocytes, 5 per cent monocytes, and 5 per cent eosinophils; urine, normal findings.

Roentgenograms.—Chest films exposed at another hospital on the day of admission showed increased radiolucency of the left hemithorax with shift of the mediastinum to the right. Examination at the University Hospital confirmed these observations (Fig. 5). In addition, the left hemidiaphragm was noted to be depressed. On fluoroscopy there was decreased motion of the depressed left hemidiaphragm. No shift of the mediastinum with respiration was noted. Twelve hours after admission additional films of the chest were exposed. There was a further increase in the radiolucency of the left hemithorax with additional shift of the mediastinum to the right. It was felt that the infant had obstructive emphysema of the left upper lobe.

Course.—During the day of admission the infant's respiratory distress increased in severity. With the confirmation by chest films of increasing emphysema of the left upper lobe it was felt that lobectomy should be performed as an emergency.

Operation.—The infant was anesthetized with ether-oxygen in a closed system. An endotracheal tube was not employed. A left thoracotomy was performed and the chest was entered through the fourth intercostal space. Inspection showed a firm, pale mass of distended lung which filled the entire pleural cavity. This could not be deflated by compression. With some difficulty the mass was delivered into the wound and was found to be the left upper lobe. The small, compressed left lower lobe could then be inflated easily by the anesthetist. The infant's condition immediately improved.

A left upper lobectomy was easily performed by the individual ligation technique. The left upper lobe bronchus was not grossly abnormal, although it could be compressed very easily. There were no enlarged lymph nodes about the bronchus. No external source of compression was found. The left lower lobe filled the pleural cavity well.

Anterior and posterior drainage tubes were inserted. The chest was closed in layers with fine silk. Blood loss was 96 c.c. The infant received 100 c.c. of whole blood during the operation. He was returned to the ward in excellent condition.

Examination of the Specimen.—No gross obstruction of the left upper lobe bronchus was found. Cartilage rings were present in the bronchial wall. There was uniform, marked emphysema of the entire lobe. On microscopic examination there was marked emphysema with dilatation of the small bronchi and bronchioles. No evidence of infection was found. Elastic tissue stains demonstrated a normal amount of elastic tissue to be present.

Fig. 5.

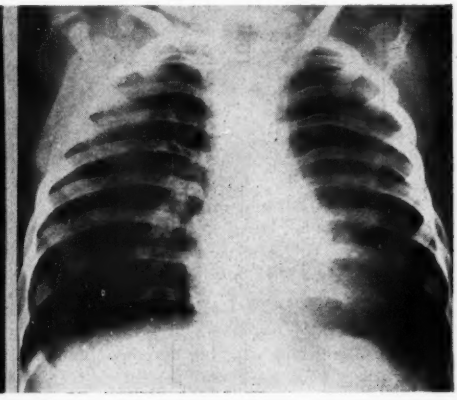

Fig. 6.

Fig. 5 (Case 2).—Preoperation roentgenogram exposed Sept. 14, 1951. There is emphysema of the left upper lobe with depression of the left hemidiaphragm and shift of the mediastinum to the right.

Fig. 6 (Case 2).—Postoperation roentgenogram exposed Feb. 9, 1953. Localized emphysema is present at the left base.

Postoperative Course.—The infant's recovery from the operation was dramatic. He had no further respiratory symptoms. The left lower lobe expanded and filled the left hemithorax. There was slight shift of the mediastinum to the left. The infant ate well and gained weight rapidly. He was discharged Sept. 25, 1951.

Course Following Discharge.—He was well for three months. At that time he began to have attacks of wheezing respirations associated with marked activity. He developed repeated upper respiratory infections usually accompanied by wheezing. On several occasions penicillin seemed to produce relief. The infant was readmitted to the University Hospital in March, 1952. At this time he had wheezing, cough, and moderate dyspnea. He was thought to have infectious tracheobronchitis. He quickly responded to antibiotics, steam inhalations,

and ephedrine and was discharged. However, despite normal growth and weight gain, he continued to have episodes of mild respiratory difficulty.

The infant was last examined Feb. 9, 1953. He had been hospitalized three times since operation for apparent respiratory infections. There had been many less severe episodes of wheezing and difficulty in breathing. The child's personal physician believed that he was sensitive to dust and bacteria and was undertaking a program of desensitization.

On examination the child was well developed and active. Breath sounds were decreased over the left chest but physical examination otherwise showed only normal findings.

Chest films exposed at this time showed mild generalized emphysema with increased emphysema at the left base (Fig. 6). There was slight compression of the left hemidiaphragm posteriorly. Fluoroscopy confirmed these findings. The diaphragms moved normally. There was no mediastinal shift with respiration.

CASE 3.—

History.—S. F., U. H. No. 725125, a 3-month-old white female infant, was admitted to the Pediatric Service of the University Hospital April 8, 1952, with a history of difficult respiration. The infant was born Dec. 27, 1951, following a normal pregnancy and an uncomplicated delivery. The infant weighed 6 pounds, 9 ounces at birth. It was thought that there was slight blueness about the mouth and questionable discoloration of the forehead shortly after birth. No other abnormalities were noted during the first seven weeks of life except that the infant nursed poorly.

Feb. 13, 1952, when she was 7 weeks old, she developed symptoms resembling a mild upper respiratory infection. Twelve hours later, while feeding, she became dyspneic and cyanotic. The infant was immediately taken to another hospital in a state of collapse. She had severe respiratory distress and was in critical condition. On admission to the hospital it was felt that she had massive collapse of the right lung. Bronchoscopy was carried out but no abnormality was found. The infant was finally resuscitated and thereafter slowly improved. After three weeks she was discharged. Ten days later wheezing respirations developed again and the infant became cyanotic. Following temporary admission to another hospital she was transferred to the University Hospital.

Family History.—Two brothers, four and five years of age, were normal. There was no family history of allergy or of congenital abnormalities.

Physical Examination.—Temperature was 100° F. (rectally); pulse, 146; respirations, 36.

The infant was well developed and was in no acute respiratory distress. However, respirations were rapid and were moderately labored. The infant's color was dusky when she cried. On examination there was hyperresonance to percussion over the left chest. Breath sounds and tactile fremitus were reduced over the left chest. The mediastinum was shifted to the right. The remainder of the physical examination showed only normal findings.

Laboratory.—Kahn test was negative; hemoglobin, 9.8 Gm.; red blood cells, 3.9 million; white blood cells, 9.400 with 43 per cent polymorphonuclear leukocytes, 55 per cent lymphocytes, and 2 per cent eosinophils; urine, normal findings.

Roentgenograms.—Chest films exposed at another hospital during the infant's original acute illness showed marked emphysema of the left upper lobe with mediastinal shift to the right. Inspiratory and expiratory films, exposed Feb. 22, 1952, were interpreted as showing emphysema of the left upper lobe with atelectasis of the left lower lobe. The heart and mediastinum were shifted to the right. These changes were more marked on expiration. It was believed that an expiratory block was present. Additional films exposed at intervals presented the same appearance.

Films exposed in the University Hospital at the time of admission confirmed these findings (Fig. 7). On fluoroscopy the mediastinum shifted further to the right on expiration. The hemidiaphragms were depressed and movement limited, particularly on the left.

Course.—The infant was not acutely ill while she was in the hospital. Because she had been in desperate straits at one time and because obstructive emphysema of the left upper lobe persisted, it was felt that lobectomy should be carried out.

Operation.—On April 16, 1952, anesthesia was induced with ether-oxygen and an endotracheal tube inserted. A left thoracotomy was carried out through the fourth intercostal space. When the pleural space was entered, there immediately presented a tense mass of air-containing lung which appeared to fill the entire hemithorax. This was found to be the left upper lobe. It could not be deflated. The small left lower lobe was collapsed but it expanded when the emphysematous left upper lobe was delivered into the wound.

A left upper lobectomy was performed employing the individual ligation technique. A probe was passed easily into the left main bronchus through the stump of the left upper lobe bronchus. No gross bronchial abnormality was found. There was no external compression of the bronchus.

Upon completion of the lobectomy it was seen that the expanded left lower lobe did not fill the pleural space and that considerable mediastinal shift would be necessary to obliterate the residual space. Anterior and posterior drainage tubes were inserted. The chest was closed with fine silk. Blood loss was 154 c.c. The infant received 210 c.c. of whole blood during the operation. The infant was returned to the ward in excellent condition.

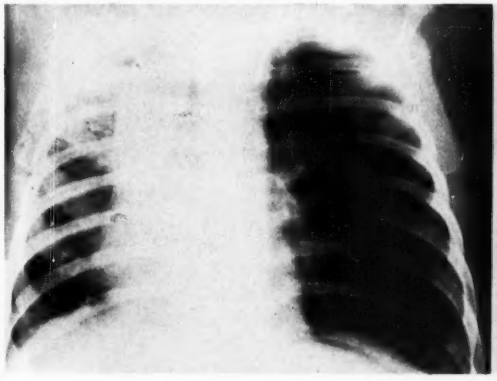

Fig. 7.

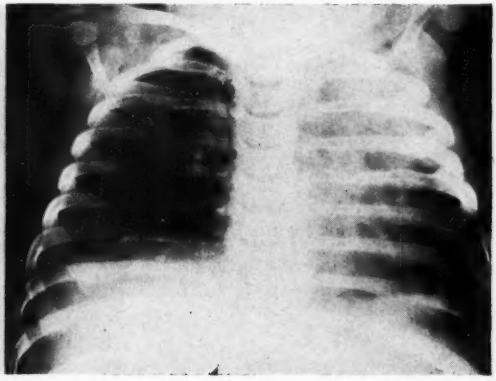

Fig. 8.

Fig. 7 (Case 3).—Preoperation roentgenogram exposed April 9, 1952. Marked emphysema of the left upper lobe is present. The left lower lobe is compressed. The mediastinum is shifted to the right, and the left hemidiaphragm is depressed.

Fig. 8 (Case 3).—Postoperation roentgenogram exposed Jan. 27, 1953. There is no emphysema. The mediastinum is shifted to the left.

Examination of the Specimen.—No gross bronchial abnormalities could be found. Bronchial cartilages were present. There was uniform, massive emphysema of the entire left upper lobe (Fig. 9). Microscopic examination confirmed the gross findings (Fig. 10). There was emphysema of the pulmonary parenchyma with dilatation of the small bronchi and bronchioles. No infection was present. Elastic tissue stains showed normal amounts of elastic tissue.

Postoperative Course.—The infant did well following operation. There were no further respiratory symptoms. The residual left pleural space was obliterated with expansion of the small left lower lobe and shift of the mediastinum to the left. The infant seemed to tire easily but eight days following operation she began to gain weight steadily. She was discharged April 27, 1952.

Course Following Discharge.—The infant was returned to the care of her personal physician following discharge. At monthly intervals she had episodes of cough and wheezing

Fig. 9 (Case 3).—Photograph of the cut surface of the left upper lobe. There is emphysema of the entire lobe.

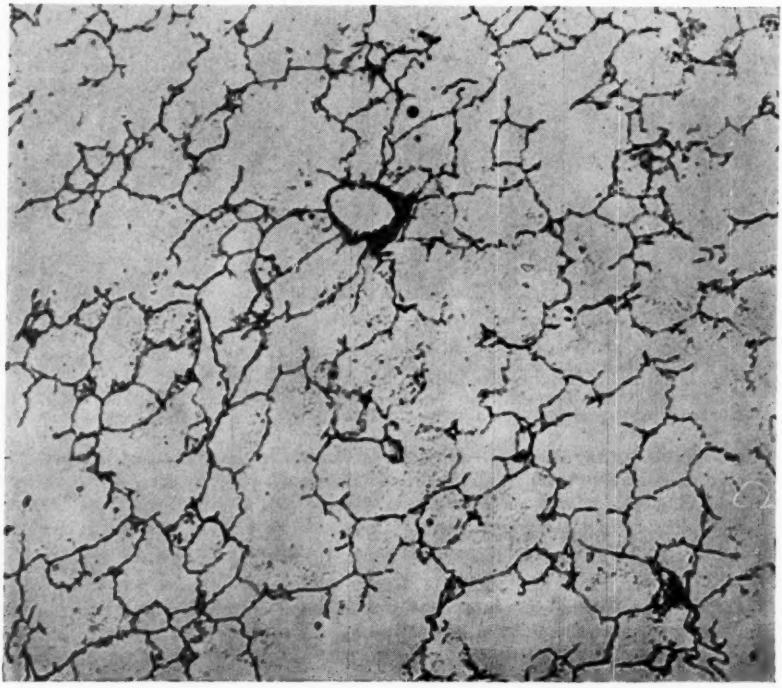

Fig. 10 (Case 3).—Photomicrograph of a section of the left upper lobe. Marked emphysema is present. There is dilatation of a small bronchus. No infection is seen (×75).

respirations which were believed to accompany respiratory infections. She was not cyanotic. Development was normal. The infant was last examined Jan. 27, 1953. At this time she was normal except for evidence on physical examination of mediastinal shift to the left.

Films exposed at this time confirmed the pronounced mediastinal shift to the left with herniation of the right lung across the anterior mediastinum (Fig. 8). On fluoroscopy there was diminished movement of the left hemithorax which seemed smaller than the right. The hemidiaphragms moved well. There was no shift of the mediastinum with respiration. There was no evidence of emphysema.

CASE 4.—

History.—F. L., U. H. No. 729545, a 2½-month-old white male infant, was admitted to the Pediatric Service of the University Hospital June 16, 1952, with a history of coughing and cyanosis. The infant was born at term March 27, 1952, following a normal labor. He weighed 7 pounds, 4 ounces at birth. The infant nursed well and was discharged home five days after birth.

Soon after he reached home it was noted that he breathed heavily. When he was 4 weeks old he began to have more difficulty breathing. He grew progressively worse. Four days after the onset of the illness the infant became cyanotic during feeding. Other, similar episodes followed. The child was admitted to another hospital where he received penicillin but did not improve. He was finally transferred to the University Hospital.

Family History.—A sister, 5½ years old, and a brother, 2½ years old, were well. There was no family history of allergy or congenital abnormalities.

Physical Examination.—Temperature, 99.2° F. (rectally); pulse, 144; respirations, 46. The infant was well developed. Respirations were rapid and labored. He was slightly cyanotic. The percussion note over the right chest was hyperresonant and breath sounds were markedly diminished. The mediastinum was shifted to the left. On ascultation of the heart there was a harsh, systolic murmur over the entire precordium. This was loudest at the apex. It radiated up the left border of the sternum. The liver edge was palpated 3 cm. below the right costal margin. The remainder of the physical examination was within normal limits.

Laboratory Data.—Kahn test was negative; hemoglobin, 10 Gm.; red blood cells, 3 million; white blood cells, 6,000 with 32 per cent polymorphonuclear leukocytes, 67 per cent lymphocytes, and 1 per cent eosinophils; urine, normal findings.

An electrocardiogram obtained June 17, 1952, showed a heart rate of 140. The P-R interval was 0.12 and the QRS, 0.07. There was slight right axis deviation, but the tracing was within normal limits for an infant of this age.

Roentgenograms.—The infant was fluoroscoped June 17, 1952. Both lungs appeared somewhat hyperaerated, although this was more marked on the right. Both hemidiaphragms were low and motion of each was restricted. There was mediastinal shift with respiration. Films exposed at this time confirmed the fluoroscopic findings (Fig. 11). These examinations were repeated June 26, 1952, and the same findings noted. It was felt that the infant had bilateral emphysema but that this was much more marked in the right middle lobe.

Bronchoscopy.—On July 2, 1952, the infant was bronchoscoped with a 3.5 mm. bronchoscope. The bronchi collapsed with each expiration. No other abnormalities were noted in the major bronchi.

Course.—During the infant's stay in the hospital he did well as long as he remained in oxygen. When he was removed from oxygen he became cyanotic, particularly when he cried. During these episodes he developed a wheezing cough and became progressively more dyspneic and cyanotic. He was relieved when oxygen was then administered. The infant was believed to have a congenital heart lesion, probably an interventricular septal defect. However, in spite of the evidence of bilateral emphysema in the roentgenograms, it was felt that the respiratory distress was due primarily to the marked emphysema of the right middle lobe.

Operation.—On July 8, 1952, a right thoracotomy was performed through the fifth intercostal space. Ether-oxygen anesthesia was administered through an endotracheal tube. As the chest was opened the greatly distended, tense right middle lobe was seen to fill the entire pleural cavity. The right upper lobe and the right lower lobe were compressed, although they contained air. The middle lobe could not be compressed. The upper and lower lobes expanded when the middle lobe was delivered into the wound.

A right middle lobectomy was carried out by the individual ligation technique. No bronchial abnormality was found. The bronchus was of normal caliber and contained cartilage rings. As the bronchus was being exposed, there was difficulty with the infant's airway. Cyanosis and cardiac slowing resulted. As soon as the airway was re-established, the infant improved. Lobectomy was then completed without further incident. The right upper lobe and the right lower lobe easily filled the pleural cavity. Anterior and posterior drainage tubes were inserted. The chest wall was closed in layers with fine silk. The blood loss was 82 c.c. The infant received 120 c.c. of whole blood. He was returned to the ward in good condition.

Fig. 11.

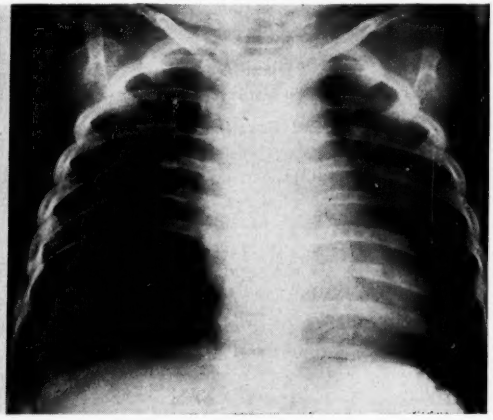

Fig. 12.

Fig. 11 (Case 4).—Preoperation roentgenogram exposed June 17, 1952. There is bilateral pulmonary emphysema which is most marked in the right middle lobe.

Fig. 12 (Case 4).—Postoperation roentgenogram exposed Jan. 15, 1953. Moderate bilateral emphysema persists. The pulmonary emphysema is most marked in the lower lobes.

Examination of the Specimen.—No gross bronchial abnormality was found. There was marked, diffuse emphysema of the entire middle lobe (Fig. 13). On section there was emphysema of the pulmonary parenchyma with dilatation of the small bronchi and bronchioles. Heavy plasma cell infiltration was seen about one bronchiole. Elastic tissue stains showed normal amounts of elastic tissue to be present.

Postoperative Course.—The infant did well following operation. He soon began to gain weight and had no further respiratory symptoms. He was not cyanotic out of oxygen. However, films of the chest exposed July 15, 1952, were still interpreted as showing evidence of emphysema of both lung fields. The infant was discharged July 19, 1952, eleven days following operation.

Course Following Discharge.—The infant did well following discharge. For a time he had rare episodes of shortness of breath and slight cyanosis. He had frequent upper respiratory infections. More recently no cyanosis has been noted. The infant was last examined

Jan. 15, 1953. Development was normal. The heart sounds had not changed. Breath sounds were decreased below the right scapula.

Fluoroscopy was carried out and again it was felt that there was a moderate degree of emphysema bilaterally, particularly in the lower lung fields. There was no mediastinal shift with respiration. Movements of each hemidiaphragm were limited. Films confirmed these findings (Fig. 12).

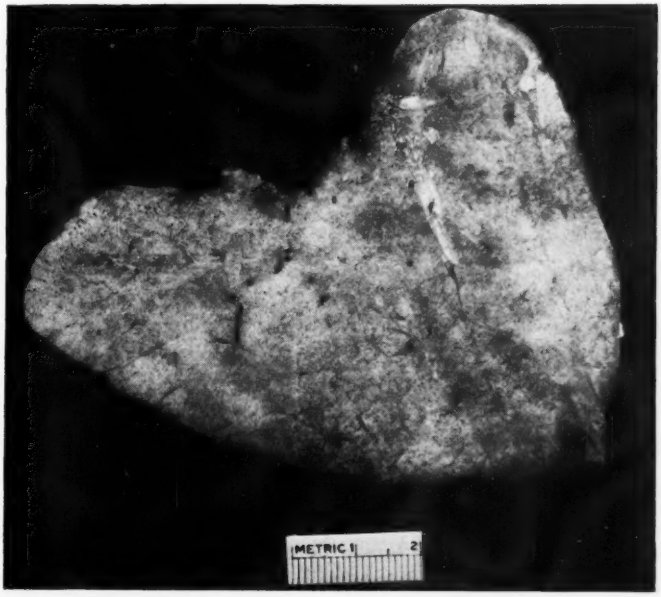

Fig. 13 (Case 4).—Photograph of the cut surface of the right middle lobe. Emphysema of the entire lobe is present (×75).

DISCUSSION

The clinical pictures presented by these four infants were strikingly uniform. Respiratory difficulties began early in infancy. The infants had labored respirations, wheezing, cough, and cyanosis. The onset of symptoms was thought most frequently to be related to a respiratory infection, but none of the infants had such an infection when examined at the time of admission. The use of antibiotics did not produce any striking improvement in their condition. On physical examination there was hyperresonance to percussion over the involved hemithorax with diminution of breath sounds on auscultation. An expiratory wheeze was often present when the infants cried.

Roentgenograms and fluoroscopy of the chest showed extensive emphysema most marked in a single lobe. This emphysematous lobe filled the entire hemithorax. There was compression of adjacent lung tissue. The mediastinum was shifted to the opposite side. The hemidiaphragm on the affected side was depressed. Shift of the mediastinum with respiration could be demonstrated

in two of the infants. In none of the infants was there clinical or roentgenographic evidence of regression of the emphysema prior to operation, and in two of the infants progression of the emphysema occurred. The emphysema was thought to be obstructive in nature.

This clinical and roentgenographic picture, once understood, was easily recognized. However, in each case there had been confusion about the diagnosis at some time during the infant's illness. Most frequently it was thought that the infant had a large pulmonary cyst. On other occasions it was felt that atelectasis or pneumonitis was present in the compressed lobes and that the emphysema was compensatory. In one infant a diagnosis of massive collapse of the opposite lung was made.

Two of the infants were bronchoscoped prior to operation. The only abnormality noted was a tendency for the bronchi to collapse during expiration. No local obstruction was seen. A third infant was bronchoscoped at another hospital and was reported to have a normal bronchial tree.

No attempt was made in any of the infants to decompress the emphysematous lobe by thoracentesis prior to operation. Such an attempt would appear to be inadvisable. In several instances in which thoracentesis was attempted by others in similar patients, a complicating tension pneumothorax occurred which made the infant's condition more precarious.[11, 14] Effective decompression of an emphysematous lobe would be difficult to accomplish by thoracentesis.

Despite careful examination of the lobes at the time of exploration and following removal, no specific bronchial obstruction producing a check valve was found in any of the specimens. Abnormal flaccidity of the bronchial walls was probably responsible for the obstructive emphysema observed. This bronchomalacia may have been the result of abnormal cartilage rings.[18] The observation that the bronchial walls collapsed on expiration in the two infants bronchoscoped before operation supports this conclusion. All of the specimens showed similar microscopic changes. There was uniform, marked emphysema with dilatation of the small bronchi and bronchioles. Elastic tissue stains showed normal amounts of elastic tissue to be present. There was little infection in the bronchi and none in the distended alveoli.

A number of etiological agents have been implicated by others in an attempt to explain the obstructive emphysema present in their patients. As causes of obstruction the following abnormalities have been listed: (1) absent or abnormally soft bronchial cartilage rings, (2) a redundant flap of bronchial mucosa, (3) compression of the bronchus by an abnormal vein, and (4) compression of the bronchus by the ductus arteriosus. However, in several of the reported cases no bronchial obstruction could be found.

The course of these infants following discharge has been in decided contrast to that reported by other authors. All of the infants did well in the immediate postoperative period; in each, the respiratory symptoms were completely relieved. Following discharge the infants remained improved and none had a return of the severe symptoms which were present before operation. Despite this improvement, each of the infants seemed more prone to develop respiratory infections and had more marked symptoms with each infection

than would have been anticipated in a normal infant. Two of the infants were admitted to the hospital several times with wheezing respirations and dyspnea. These symptoms appeared to accompany a respiratory infection. Sensitivity studies on two of the infants disclosed no specific allergies.

Roentgenograms of two of the infants following operation showed emphysema in other areas of the lung fields. This has persisted without significant change. The emphysema was felt to be more than could be attributed to compensatory emphysema following expansion of collapsed pulmonary tissue after operation. In a third infant there was evidence of mild generalized emphysema during a respiratory infection.

It would appear that the bronchial abnormality which produced the primary lobar obstruction involved other bronchi and, perhaps, the entire bronchial tree to a lesser degree. The symptoms which these infants had following operation may have been due to flaccidity of other portions of the bronchial tree. The tendency toward obstruction during expiration probably was accentuated by respiratory infections.

SUMMARY

1. Severe obstructive emphysema, most marked in a single lobe, has been observed in four infants. In two of the infants the left upper lobe was involved; in two, the right middle lobe.

2. Lobectomy was performed successfully in each of the infants, the youngest having been 6 days old and the oldest 8 months old.

3. Each of the lobes removed showed marked, generalized emphysema. Infection was not present. No specific bronchial obstruction was found. It is believed that the obstructive emphysema present was due to a check valve mechanism produced by abnormal flaccidity of the bronchi.

4. Each of the infants was improved by lobectomy. However, all had respiratory symptoms following discharge. In two of the infants there was roentgenographic evidence of emphysema in other portions of the lungs following operation. It is suggested that the bronchial abnormality also involved other portions of the bronchial tree.

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DISCUSSION

DR. PAUL C. SAMSON, Oakland, Calif.—Dr. Sloan's paper is most interesting, and I would like to discuss it, not because it involves infants but perhaps because the question of congenital emphysema is under discussion.

I would like to report a case involving a young man who came to me primarily because of a pectus excavatum (and in that connection perhaps Dr. Shaw will say something). There may be a connection between congenital lobar emphysema and pectus excavatum; we do not know.

(Slide.) This patient, aged 21, showed on his x-ray pictures a large area in which the lung markings were relatively meager and we thought this was some type of cystic disease.

(Slide.) This shows the marked pectus, and again that there is lack of normal pulmonary markings in the right mid-lung field. We decided to do an exploratory operation first, and performed a right thoracotomy.

(Slide.) Much to our surprise we found an enormous middle lobe which went from apex to base and, as you see, even out of the chest. The lobe is tremendously blown up. Grossly and microscopically this was congenital emphysema insofar as we could tell, because of the fact that there was diffuse widening of all the alveolar spaces and relative lack of bronchial elements.

(Slide.) Six months later we repaired the pectus with a strut, after the manner of Dailey, using a bone bank bone, and the patient has remained well. I have no words of wisdom on this, but it is of interest in that the patient had both a pectus and apparently congenital localized emphysema of one lobe.

DR. ROBERT R. SHAW, Dallas.—I have enjoyed this paper by Dr. Sloan, largely because of the fine manner in which he has presented the problem and also because of the interest I have had in this condition for several years. I would like to present two cases to help point out the differentiation between this condition and the usual type of obstructive emphysema that we see with thick viscid mucus in the bronchi of an infant.

(Slide.) This case is very similar to the first case shown by Dr. Sloan. The child was three months old and the only symptom was rapid respirations and wheezing. There had been no evidence of infection at any time and the child was developing well. The diagnosis of emphysema on the right had to be made by a correlation of the physical findings of no breath sounds on the right, with very good breath sounds on the left, which seems a paradox when one looks at the films of the chest.

(Slide). This shows the right middle lobe removed from this 3-month-old infant. It filled the right hemithorax entirely, compressing the upper and middle lobes which were not air-containing. The child has done well since surgery and has had no further respiratory difficulty. She is now a little over 2 years old.

(Slide.) Here we had an opportunity to observe the permanence of this situation. The slide on the left shows a child 2 weeks of age. At that time it was thought that an enlarged thymus was causing the obstructive type of breathing and x-ray therapy was given. It was not noticed that there was a translucency in the upper part of the left lung field. The second slide shows an x-ray picture taken when the child was 2 years old, and the same translucency is present with the heart still shifted into the right chest.

(Slide.) On the left is shown the child at the age of 4, so there had been an interval of two years between these x-ray views, and there is still translucency on the left, persisting in proportion to the child's growth. I found that the apical posterior segment of the left upper lobe was involved and was easily stripped away from the remaining portion of the lobe. Because of a severe pectus excavatum which had developed during the four years of growth, the heart was unable to go back into the left chest. Surgery for this condition was advised but the parents were so pleased with the respiratory improvement that nothing further has been done.

(Slide.) I think it is very important to differentiate this condition from the temporarily plugged bronchus which can also cause an emphysema in infants. I am wondering how Dr. Sloan was able to differentiate in the 6-day-old child these two different pathologic entities. Here is a child 2 days old with apparent emphysema of the left lung. The child was put in moist vapor and observed for one week.

(Slide.) This shows the chest at the end of one week of treatment. The emphysema has completely cleared. Just this past week I saw a child with atresia of the esophagus, who also had emphysema of the left lung. The emphysema has completely cleared following repair of the atresia. A temporary situation due to a thick viscid mucus plug causing an obstructive phenomenon must be differentiated from the more permanent type of condition that we have called localized hypertrophic emphysema.

DR. JOHN S. HARTER, Louisville.—We have had five of these infants in the past seven years. One reason for the large number is the fact that one family produced two of them. Three of the cases are of interest.

(Slide.) This 5-month-old baby was admitted to the hospital on Dec. 20, 1949, with a history of progressive dyspnea for three weeks. X-ray examination of the chest revealed a cyst occupying the entire right pleural cavity and displacing the heart and mediastinal structures to the left. There is trabeculation throughout the right lung field typical of emphysema. Because the child was admitted about 2:00 A.M. after a trip from the southern part of the state, a catheter was inserted and it was thought that operation would have to be performed later. However, the normal lung expanded and no further operative procedure was necessary.

(Slide.) The follow-up of this patient in 1952, four years after treatment by aspiration alone, revealed the child to be well with no recurrence of the cyst. This is shown because of the possibility of treating these patients by that method.

The second patient was 23 months of age and was admitted to the hospital on May 17, 1950. The parents gave a history of the child having had pneumonia three weeks previously. During the ten days before admission the baby became increasingly dyspneic.

(Slide.) X-ray views of the chest revealed a cyst occupying the right pleural space, displacing the heart and mediastinal structures to the left. At thoracotomy the right middle lobe was found to be greatly emphysematous, filling the entire right chest. A right middle lobectomy was performed. What was thought to be an incidental finding at the time of operation, was a small fibrin ball free in the pleural cavity. Thorough search of the entire pleural cavity failed to reveal any other pathologic change.

(Slide.) Much to our surprise, this photomicrograph of the fibrin ball was interpreted as sarcoma. The next slide is a high-power photograph of the fibrin ball.

The baby made an uneventful recovery and was discharged from the hospital after two weeks, with a small amount of pleural fluid present. At six weeks a checkup showed considerable fluid in the right chest. Aspiration of the fluid again revealed tumor cells. We told the family of the fatal prognosis but they refused to believe it and took the child else-

where, where it was operated upon and died three days after operation, at which time a large sarcoma of the chest was found.

The third patient was a sister of this child, who at 10 days of age was admitted to the hospital, extremely dyspneic.

(Slide.) This was also a right middle lobe emphysema and a right middle lobe lobectomy was done. This child has remained well to date.

The last two patients were sisters and one of them died of sarcoma following operation.

The first patient presented, with the catheter drainage only, suggests that the etiology of obstructive unilobar emphysema may in some instances at least be a failure of the remaining lobe or lobes to expand normally following birth. Certainly this child has remained perfectly normal for four years. The two sisters with the emphysema of the same lobe, one with sarcoma of undetermined origin, suggest the possibility of some congenital anomaly.

DR. WILLIAM A. JOHNS, Richmond.—I wish to thank the Association for allowing me as a guest the privilege of the floor. We have had the opportunity of seeing one patient similar to the ones reported by Dr. Sloan. It is interesting to note in the literature on the subject that all cases not treated by surgical excision died.

Our case was that of a 9-year-old girl whose chief complaint was frequent episodes of vomiting and nervousness. (Slide.) The anteroposterior view of the chest preoperatively shows the heart and mediastinal structures shifted to the left, the lesion being in the left upper lobe. The lower lobe is compressed.

(Slide.) The roentgenogram and photograph of the removed specimen, taken about one hour after removal, show that the lobe is not collapsed and has the appearance of Gelfoam, as described by Shaw. The apical segment was not involved in the process.

(Slide.) These are the low- and high-power photomicrographs of the cut specimen. The alveoli are dilated and the walls are thin. The walls of the bronchioles are thin and the lumina are dilated. The lining cells consist of respiratory epithelium.

(Slide.) Here is the postoperative view of the chest one month after operation, showing that the heart and mediastinum have returned to normal position, and the left lower lobe has almost completely filled the thoracic cage. No definite obstruction in the bronchi could be found. The patient has done exceedingly well since operation.

DR. RICHARD H. SWEET, Boston.—I hesitate to enter upon this discussion but it seems to me that it would be of interest to come back to the fundamental question of etiology. I would be interested also to know what Dr. Sloan says about Dr. Harter's cases, because I do not believe he is talking about the same disease.

(Slide.) Here is an inspiration-expiration film showing the paradoxical motion on the left side of a young boy of 17. He had a lot of dyspnea on exertion. The same thing is shown in the lateral view.

(Slide.) This is the slide I wanted to show principally because we were able to show in this patient that the entire lung was involved. The bronchial cartilages are completely deficient. There was no single cartilage which had the usual U-shaped arch in continuity. The bronchial wall was soft and compressible, just as much so anteriorly as posteriorly, and I feel certain that this enormous overdistention and emphysema was due to the fact that the bronchus had no stability and performed in this abnormal manner because of lack of adequate cartilage.

DR. ROSS ROBERTSON, Vancouver.—I think it is a great compliment to the excellence of Dr. Sloan's paper that it has stirred up so much discussion. I am particularly interested in this subject.

(Slide.) This is a photograph of an autopsy performed on a child at the Sick Children's Hospital in Toronto in 1933. This child at the age of two weeks had suffered from shortness of breath and cyanosis. The right chest had been aspirated repeatedly in the belief that a cystic lung was present, but the child died. This autopsy picture shows very well the huge right upper lobe almost filling the chest, compressing the lower lobe down onto the diaphragm and compressing the left lung into the left gutter.

(Slide.) This shows a section of the right upper lobe bronchus. The autopsy was performed by Dr. Erb, the pathologist, and he came to the conclusion that this flap of mucosa which is directed distally formed a ball-valve obstruction and caused the emphysema.

(Slide.) This is an x-ray view of a patient 2 weeks old who had symptoms similar to the other patient described. You will note that the lower lobe is compressed down against the diaphragm. You can differentiate this from a traumatic surgical emphysema and hemothorax by the fact that the lung is compressed downward instead of into a ball at the hilus. It can also be differentiated from atelectasis of the other lung by the fact that the diaphragm is lower; and from cystic disease in that you can see the lung markings faintly, and there is no evidence of cyst.

Caffey has suggested that regional obstructive emphysema is usually lobar rather than segmental in distribution because of the alveolar pores of Kohn which form communicating channels between the lobar segments. Consequently, if a segmental bronchus were obstructed by a congenital check-valve, symptoms would not occur unless the pores of Kohn were obstructed by some inflammatory process. From this reason uncomplicated emphysema is lobar in distribution.

DR. J. L. EHRENHAFT, Iowa City.—In the last eighteen months we have encountered six newborns and infants with the condition under discussion; I would like to show some slides.

(Slide.) The first slide shows the preoperative chest x-ray photograph of a 5-month-old infant who has, as you will see later, congenital cystic disease involving the left upper lobe of the lung. You see the marked displacement of the mediastinum to the right with compression of the contralateral lung. This child showed extreme respiratory distress when first seen by us.

(Slide.) This slide shows the operative procedure in progress. It demonstrates the greatly enlarged left upper lobe and the compression of the normal left lower lobe. The left upper lobe, particularly the anterior pectoral segment and the subapical segments were involved and showed multiple smaller and larger cystic structures.

(Slide.) This is a photomicrograph of the tissue removed by segmental resection, demonstrating that this lesion was not obstructive emphysema but true congenital cystic disease.

I would like to show you also the chest x-ray films of another patient, 12 days of age. This child also was extremely ill when seen by us. If you compare the two chest x-ray films on the slide you will see marked progression of the area of emphysema in the right hemithorax. The time between the two chest x-ray pictures is twelve hours. The pathologic examination after the markedly emphysematous right middle lobe had been resected revealed it to be true emphysema, probably obstructive in type. These children are extremely ill and die a progressive anoxic death unless rapid surgical intervention is available.

I would like to inject a word of caution. We have observed that the interval between preoperative intratracheal intubation for anesthesia purposes and the time of opening the chest is an extremely critical one. Apparently during positive pressure anesthesia, further rapid expansion of the emphysematous area does occur. This is immediately relieved as soon as the thoracic cage is opened and the remainder of the lung tissue is permitted to expand. In the patients we have seen, the history of forceful resuscitative measures in the post-partum period has been elicited. Therefore, we became interested to see if some of the emphysematous damage may be due to resuscitative measures.

I would like to show you two slides demonstrating these points. The first one is from a newborn infant who died two hours after birth. It had forceful resuscitative measures and it demonstrates the marked emphysematous changes in the lung parenchyma. In the second instance, you will see the microscopic appearance on the slide. Forceful resuscitative measures were also employed. It demonstrates marked interstitial emphysema and areas of unexpanded lung tissue. We are convinced that irreparable damage can be caused in the immediate post-partum period by overforceful resuscitative measures.

DR. WILLIAM HENRY, Lynwood, Calif.—I too would like to thank the Association for the privilege of discussing Dr. Sloan's paper. I have had experience with one patient, a 10-year-old boy with emphysema localized to three separate areas of the right lung. One was in the right upper lobe, two in the right lower lobe.

(Slide.) This is the chest film. At this time it was thought that the patient had obstructive emphysema due to an intrabronchial foreign body. Bronchoscopy was carried out and no abnormality was found. Following a bronchogram it was thought that he had cystic disease of the lung and a right thoracotomy was performed.

(Slide.) This is the artist's conception of the operative findings. As nearly as I could determine the emphysema involved the posterior segment of the right upper lobe, and the lateral basal and superior segments of the right lower lobe. At that time, a little over a year ago, I did not know much about this disease. Resection of the three areas was carried out from the periphery inward. This was done very easily (not that I would recommend this method of resection in every case) as in this particular type of disease the line of demarcation with the normal lung is extremely simple to follow.

(Slide.) This photomicrograph shows the emphysema, and the next slide is a postoperative film showing the patient three months later. He has been followed for more than one year and has no symptoms whatsoever.

DR. LEO ELOESSER, San Francisco.—This extremely varied series of most interesting reports on infantile respiratory difficulties indicates great possibilities. The reports range from bronchomalacia to tumors, to adenopathy, to cysts, and to a most varied series of pathologic conditions. When I was younger they would have fallen into that undiagnosed limbo of diseases known as croup. I wonder whether Dr. Johns means that the mortality of infantile croup is 100 per cent, because, as I understand it, there are many patients with croup who recover completely. It indicates, I think, the need for more exact diagnostic procedures, and it indicates also the possibilities of being more exact in our diagnosis before we proceed to resection. In some of these infants, especially those with a very large congenital cyst, surgical procedure was evidently indicated. In some of them, perhaps, I wonder whether the procedure was necessary, or whether they might have recovered without losing one-half or one-quarter of their lungs, and without being headed for the scoliosis which some of the x-rays already lead one to suspect.

DR. SLOAN (Closing).—I should like to thank the discussors and answer the questions which were raised. In reply to Dr. Shaw, it was not known whether the 6-day-old infant presented had permanent changes in the emphysematous left upper lobe and whether he might not have recovered without operation. Had more expert care been employed and had bronchodilators been used more effectively, he might have been tided over. Operation was performed because it was thought the child was going to die.

I agree with Dr. Sweet that this condition should be differentiated from the large pulmonary cysts which some of the discussors have mentioned. It should also be pointed out that thoracentesis for decompression of an emphysematous lobe may be fraught with considerable danger, as previous reports indicate.

In contrast to Dr. Ehrenhaft's patients, none of the infants presented here required vigorous resuscitative measures at birth. While too vigorous resuscitative measures are an important factor in certain respiratory emergencies at birth, they played no part in the obstructive emphysema observed in these infants.

MUCOCELE, CONGENITAL BRONCHIECTASIS, AND BRONCHIOGENIC CYST

BEATTY H. RAMSAY, M.D., AND FRANCIS X. BYRON, M.D.
LOS ANGELES, CALIF.

THE LITERATURE on cystic changes in the lungs is very extensive. Several classifications and theories of etiology have been presented. There have been recent reviews of the subject, including an excellent paper by Cooke and Blades.[1] In the literature, we have been unable to find a recorded case of mucocele of the lung or a specific reference to the presence of bronchial septa or segments of atresia in the bronchiogenic pulmonary cysts. During the past three years, we have encountered three cases of pulmonary cystic change with coexistent proximal bronchial septal obstruction. We do not believe this occurrence is unusual and coincidental. We do believe that bronchial obstruction, usually due to a congenital septum, is the common cause of pulmonary bronchiogenic cysts.

The first case, one of a mucocele of the lung, has been reported.[2] The patient was a 17-year-old-girl with pulmonary tuberculosis, and x-ray examination showed a single cystic lesion in the left lower lobe. At surgery, the findings included in Case 1 were evident:

CASE 1.—M. S. was admitted to the Orange County General Hospital July 22, 1950. There were filmy adhesions between the superior and posterior segments and the adjacent chest wall. The arrangement of the lung was abnormal in the following respects: The basal segments were divided into two equal portions by a long vertical fissure; there was a partial fissure between the superior segment of the lower lobe and the basal segments; the interlobar fissure was incomplete, being totally absent in the posterior half; the bronchi of the posterior segment of the upper lobe originated from a subsegmental bronchus (most posterior) of the superior segment of the lower lobe, and thus traversed the posterior portion of the usual fissure line.

Dissection revealed smooth, rounded, pale, semitranslucent, cystlike structures, extending through the superior and posterior segments in a branching, budding fashion. When an occasional branch was ruptured during the dissection, it was found to contain a large amount of thick, whitish, mucoid material. There was absolutely no evidence of inflammation or of solid deposit.

After division of the superior segmental bronchus of the lower lobe, it was possible to visualize the origin. This was imperforate, coming to a shallow, blind, funnel-shaped pocket, the walls of which were completely smooth as were the walls of all the cystic spaces.

The entire lung had relatively little carbon deposit, but the superior segment was totally devoid of such pigmentation. All segments of the lung seemed to inflate normally, although the posterior segment deflated somewhat more slowly than the other. The blood supply, artery and veins, and the remaining hilar and pulmonary anatomy were normal. The diaphragm, pericardium, and mediastinal structures also were normal.

Read at the Thirty-third Annual Meeting of The American Association for Thoracic Surgery, San Francisco, Calif., March 27, 28, and 30, 1953.

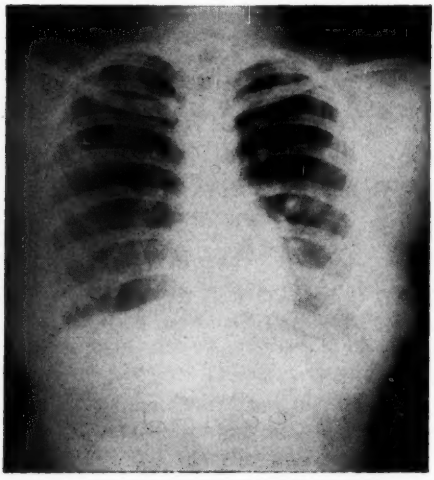

Fig. 1 (Case 1).—Posteroanterior roentgenogram. There is a circumscribed density in the left mid-lung field. An air-fluid level is present. A multi-branched density cannot be seen.

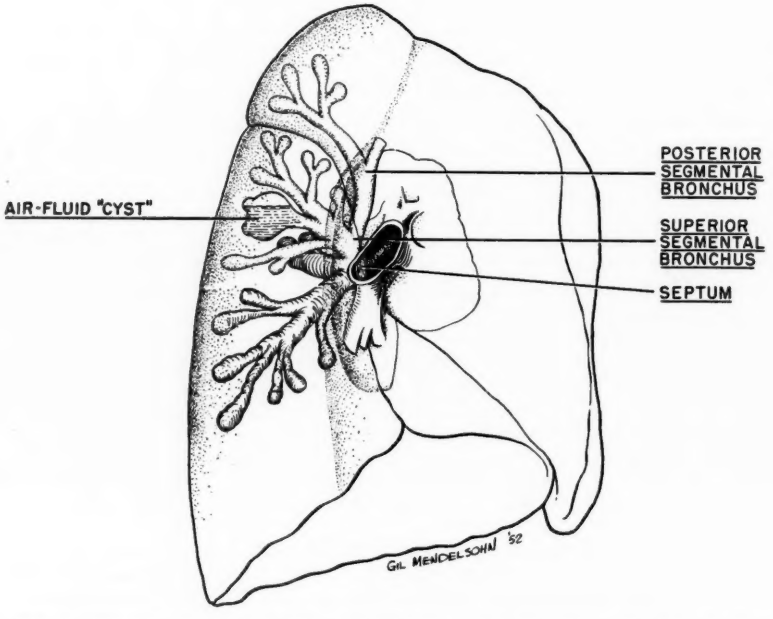

Fig. 2.—Mucocele of the left lung. Illustration of the bronchial pathology in Case 1. The lower lobe superior segmental orifice is obstructed by a flush thin membrane. The bronchi distal to this septum are dilated and filled with mucus. One such bronchus is cystic and contains both mucus and air. The upper lobe posterior segmental bronchus terminates aberrantly distal to the septum. Both obstructed segments are normally inflated.

Surgical removal was by segmental resection. The area of bronchial obstruction was not removed. The septum was flush with the superior segmental orifice, as demonstrated by bronchoscopy, and was not removed. This septum was thin and was funnel shaped from the distal side, as one would expect from the tension of the mucus within the dilated bronchial branches.

The second occurrence was in a 30-year-old woman, asymptomatic, in whom a left pulmonary mass was discovered by survey chest roentgen-ray examination. At surgery, the following pathology was demonstrated:

CASE 2.—On April 5, 1951, R. B. was admitted to Saint John's Hospital. The pleural space was everywhere free from adhesions. The lower lobe of the lung was perfectly normal. The heart and mediastinum were perfectly normal. At the hilum of the upper lobe anteriorly, there was a cystic structure with an opaque, white wall of moderate thickness. This extended from the mediastinum outward into the lung tissue between the superior segment of the lingula and the apical segment of the upper lobe. Dissection revealed that the structure was cystic and originated from the upper lobe bronchus near its origin; at this point, there was an imperforate condition. The cystic structure itself measured about 4 cm. in diameter and split the superior pulmonary vein into two parts, one of which coursed along the inferior surface and the other along the superior surface. The pulmonary artery to the left lung also was closely opposed above and slightly posteriorly. The margins of the cyst seemed to extend right to intersegmental planes above and below, so that intersegmental veins could be followed in the separation from the superior lingular segment inferiorly, and from the remaining portion of the anterior segment above.

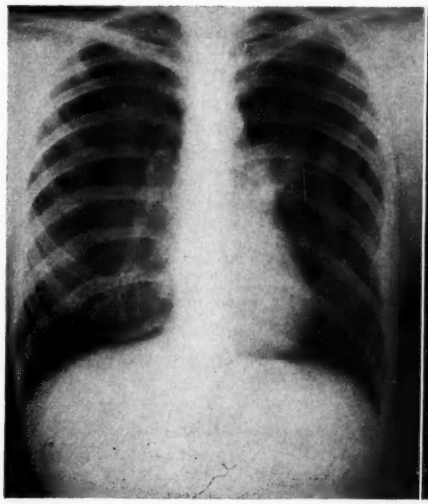

Fig. 3.

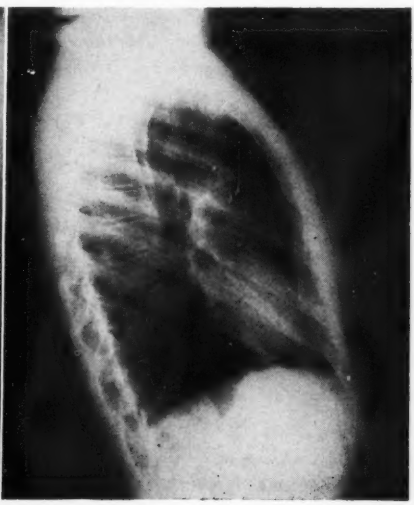

Fig. 4.

Fig. 3 (Case 2).—Posteroanterior roentgenogram. There is a circumscribed left parahilar shadow of moderate density.

Fig. 4 (Case 2).—Lateral view of the circumscribed density.

There was also an anthracotic scar near the tip of the cyst and at the lower intersegmental plane. In cutting through this a small, yellowish nodule was found, resembling a small tuberculous focus, encapsulated, and measuring about 2 mm. in diameter.

The cystic structure contained a mucoid-like material and resembled, in every respect, an obstructed, small segmental bronchus filled with mucous secretions in all its branches, as it had several small, clublike outpocketings from the main cystic mass.

An excerpt from the operative description is pertinent. "However, the stump was immediately clamped distally and was sectioned proximally, and an imperforate condition of the obstructed bronchus was seen. This was perfectly airtight and it was not necessary to place any sutures here. The cyst was then removed from the upper lobe by following small veins as a guide to the upper and lower intersegmental surfaces."

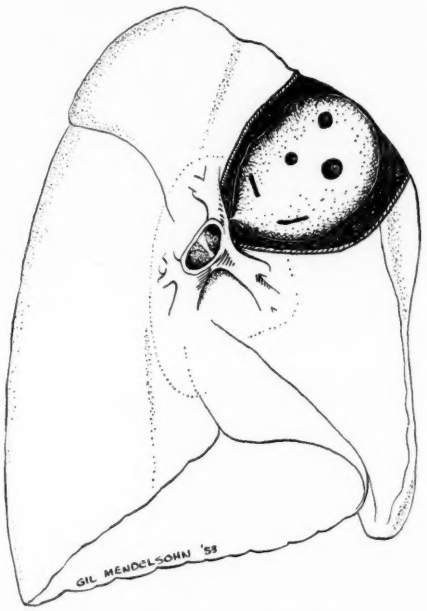

Fig. 5.—Bronchiogenic cyst left upper lobe. Illustration of the left lung depicting the cystic structure which replaces the anterior segment of the upper lobe. Note the narrow imperforate origin from the upper lobe bronchus. There are several shallow outpouchings in the cyst wall which are probably the dilated terminations of small bronchi. The wall is thickened. The intersegmental planes contain dark pigment.

The third patient was a 41-year-old man with an infected multilocular cystic mass in the middle lobe area.

CASE 3.—On Feb. 2, 1952, A. G. was admitted to Saint John's Hospital. At surgery, segmental resection of the medial segment of the middle lobe was carried out. No septum or bronchial obstruction was noted at the point of bronchial division.

Subsequently, the gross specimen was carefully examined by dissection along the bronchi. The medial segmental bronchus was normal in every respect, measured about 1 cm. in length, and bifurcated at that point. One subsegmental bronchus was dilated and thick-walled in all its branches, and at its commencement there was a rimlike circumferential thickening suggestive of residual bronchial membrane. The other subsegmental bronchus was almost completely obstructed at its origin by a thin membrane, one margin of which was in continuity with the thickening of the other subsegmental orifice. There was marked cystic change beyond the membrane, multilocular, and with varying sized out-

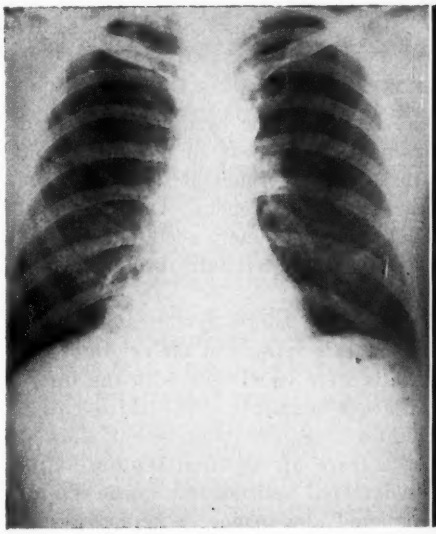

Fig. 6.

Fig. 7.

Fig. 6 (Case 3).—Posteroanterior roentgenogram. Multilocular cystic density in right cardiophrenic angle. Two fluid levels are present.

Fig. 7 (Case 3).—Lateral view again showing the two fluid levels.

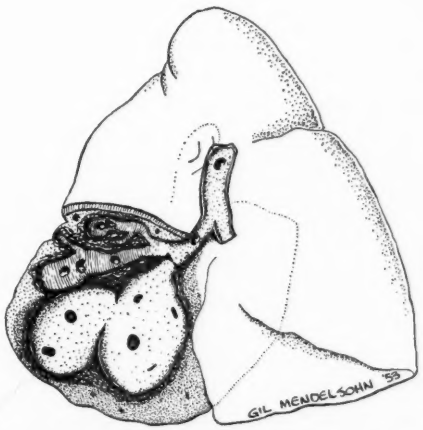

Fig. 8.—Bronchiogenic cyst and congenital bronchiectasis right middle lobe. Illustration of the right lung. In cross section the middle lobe bronchus arises from the intermediate bronchus and divides into a lateral segmental bronchus (orifice shown) and a medial segmental bronchus. The medial segmental bronchus is normally patent but soon bifurcates into subsegmental bronchi. At the bifurcation, there is an abnormal septum.

This septum almost completely obstructs one branch, and beyond it there is multilocular cystic dilatation. The only residuum of the septum across the other branch is a rimlike circumferential thickening. Beyond this the tributary bronchi show bronchiectatic changes with transverse ridging of the mucosa, thickened walls, and dilated lumens.

The parenchyma is condensed, more so about the cyst.

pouchings. There was a 2 mm. opening in the septum, through which the cystic structure communicated with the medial segmental bronchus.

The pathologic diagnosis was bronchiectasis and cystic disease of the lung.

DISCUSSION

The three cases presented (mucocele, bronchiogenic cyst, and combined congenital bronchiectasis and bronchiogenic cyst) appear to be variants of the same pathologic process. Each features a thin bronchial septum, intact or ruptured, along with distention in the tributary bronchi. One case presented a smooth, thin, translucent septum which completely occluded the left lower lobe superior segmental orifice; as a result, the tributary bronchial tree was distended with clear mucus.

The pathology of a second patient revealed a single cystic cavity, with outpouchings in the wall, and a thick, imperforate septum at the origin of the cyst from the bronchial tree; the cyst walls were coexistent with the intersegmental planes of separation from adjacent segments. The third entity involved the medial segment of the middle lobe; one subsegment showed severe cylindric bronchiectasis with just a trace of ruptured septum at its orifice; the other subsegment revealed an infected multilocular cystic dilatation beyond a thin septum which obstructed the orifice, a pinhead-sized opening being present in the septum.

Only in Case 3 was it practical to remove the septum as part of the resected specimen. Mistakenly the cyst, but not the septum, was photographed and studied microscopically. The portion of the specimen containing the septum was discarded. It is regretted that a photograph and a microphotograph of the septum cannot be submitted.

Although bronchial septa have not been described in connection with bronchiogenic cysts, there have been reports of numerous cases where the cystic spaces did not communicate with a bronchus. In Cooke and Blades' recent paper, two of the reported cases showed no filling of the involved segments on bronchography, and there was no record of bronchoscopic study. Perhaps a segment of bronchial atresia was present in each case. Where, in other reports, communication of the cystic spaces with the bronchus was shown, it is possible that a septum had originally been present, but had broken down. In the past, most cysts have been removed by lobectomy, and any membrane at a segmental orifice passed unnoticed. Likewise, pathologic study has usually been by cross-section technique rather than by dissection along the bronchial tree. Where resection has left a septum with the proximal stump (Case 1), it has been unrecorded.

Study of most illustrated cases of bronchiogenic cyst reveals outpouchings in the cyst walls. It is probable that these are the blind, dilated ends of tributary bronchi which have been taken up in the wall of the progressively enlarging cyst. So-called multiple cysts have been illustrated in cross-section view. It is doubtful that most of these are actually separate cysts. More likely, they are various-sized bronchial branches, dilated, distended, and tributary to a septal obstruction, and which appear to be separate cysts when

cut in cross section. Probably examination of such cases by bronchial study will reveal the singleness of the entity.

Of all the theories which have been advanced to explain the formation of bronchiogenic cysts, the one which seems most reasonable, in light of the findings reported here, is that of Mueller, as interpreted by Sante.[3] This theory holds that at a certain stage in bronchial development, the normal tubular growth is temporarily interrupted, and subsequently resumed. As a result, a zone of atresia is produced, short or long, distal to which bronchial tributaries exist. This theory and the circumstances of the clinical cases fit exactly. That portion of Mueller's theory which applies to alveolar-type cysts is not relevant to this paper.

The free and easy inflation and deflation of the two obstructed segments of Case 1 explains the lack of atelectasis and secondary x-ray change. It is probable that a mucocele is not evident on a roentgenograph until it becomes of sufficient mass to produce a significant density in relation to the amount of lung parenchyma in which it is found. Secondary changes of decreased lung volume or mediastinal shift do not occur until collateral ventilation is eliminated by edema, inflammation, or other change. Thus it would seem that a congenital bronchiogenic cyst, or its precursor, can be present in the total absence of roentgenographic change.

The deposition of anthracotic pigment in the adjacent intersegmental planes of Case 2 is probably a consequence of collateral ventilation and the filtering out of the particles by the fine mesh of the collateral openings.

In Case 1 there was a single thin-walled cystic structure, visible by x-ray examination. This contained both air and fluid. Yet at surgery, there was total bronchial obstruction and filling of the tributary bronchi with mucus. During segmental resection, unavoidably, small bronchi were interrupted and it was not possible to demonstrate the air-fluid cystic space. It must be assumed that the air entered this part of the mucocele from the alveoli, and not along the bronchial tree. It is of interest that the retained mucus does not enter the alveoli, probably due to physical properties of the mucus. Loosli, Adams, and Thornton[4] have produced an analogous condition in dogs. By painting a lobar bronchus with silver nitrate, aseptic bronchostenosis was produced. Since no single segment of the lobe remained unobstructed, there was no opportunity for collateral ventilation, and atelectasis resulted. On pathologic examination, the larger bronchi were distended with mucus, but the terminal bronchioles and alveoli were collapsed and free from mucus.

SUMMARY

It appears clear that during bronchial development, temporary interruption of tubular growth results in formation of an atresic area beyond which normal bronchial structure exists. Progressive accumulation of secreted mucus produces cylindric distension of the blocked bronchial branches, a mucocele. If the atresic area breaks down at an early phase of distention, true congenital bronchiectasis results. If extreme distention occurs before septal rupture, there is cystic dilatation, unilocular or multilocular, and the walls of such have

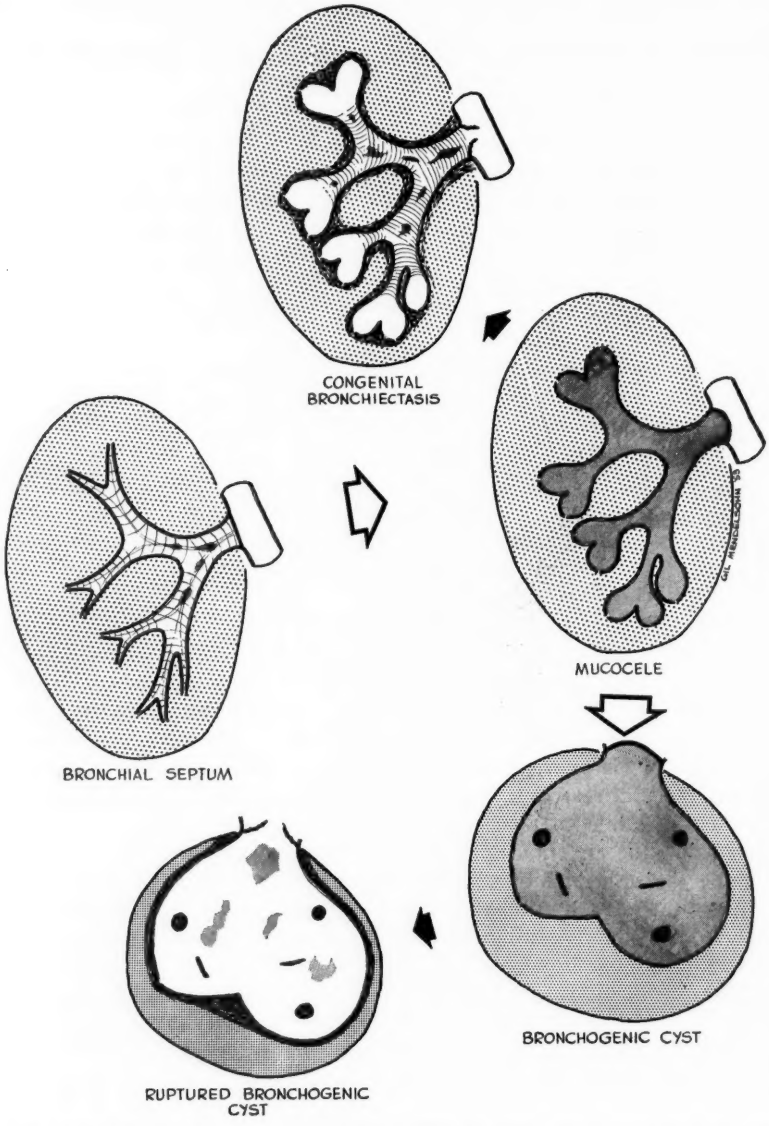

Fig. 9.—Congenital bronchial atresia, relationship of associated clinical entities. Schematic drawing. Commencing center left, follow the arrow:

Bronchial atresia: Proximally there is a segment of bronchus which failed to undergo tubular development. This area may be thin and membranous, as depicted, or longer and cordlike. Distally the bronchial wall is normal and contains functioning glands. Mucus drops are shown within the lumen. The parenchyma is normal.

Mucocele: The mucus has accumulated because of the obstruction. The septum is now convex and the bronchial walls are dilated. The parenchyma is normal.

Depending upon whether or not the septum ruptures, the following develops:

Congenital bronchiectasis: The septum of the mucocele has ruptured. Some mucus drops are still within the dilated bronchi. Infection has resulted in thickened walls.

Bronchiogenic cyst: The septum of the mucocele has remained intact and cystic dilatation of the bronchial walls has taken place. The dilated terminations of the smallest bronchi are seen as outpouchings in the cyst wall. The parenchyma is condensed but may still inflate and deflate via collateral ventilation if there are adjacent normal segments.

Ruptured bronchiogenic cyst: After cystic dilatation, the septum has ruptured. Most of the mucus has escaped. Infection has occurred with thickening of the cyst wall. The parenchyma is affected and airless.

small blind outpouchings which are the dilated terminations of the smaller bronchi.

Other findings of interest include: (1) normal inflation—deflation of the alveoli belonging to the obstructed segments, (2) the presence of an air-fluid level in one area of bronchial dilatation, and (3) failure of repeated x-ray examinations to demonstrate the branching mucocele.

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DISCUSSION

DR. HENRY J. HEIMLICH, New York.—I wish to thank the Association for the privilege of the floor. I would like to refer to a case involving a patient with a congenital intrapulmonary cyst.

This child was 34 months old at the time of admission. His mother stated that at the age of one year he was admitted to a hospital with the diagnosis of pneumonia. At that time no chest x-ray picture was taken. From the age of one year until the patient's admission 22 months later, he had recurrent bouts of fever, with temperatures of 104° to 105° F., lasting one week to ten days. He always appeared undernourished and refused food after a few mouthfuls. He could walk less than a block before he had to be carried, due to extreme fatigue. He had frequent colds and cough with expectoration. Ten days before admission he had a recurrence of the fever already described. The family physician took a chest x-ray film which showed a huge collection of fluid with a fluid level in the right chest; he thought it to be an empyema.

(Slide.) These are the admission x-ray films showing a huge cyst in the right chest. The third x-ray picture shows the postoperative film.

On admission temperature was 104° F.; he was given antibiotics for five days after which the temperature diminished to 101° F. At this time a thoracotomy was performed through an anterolateral incision because of the possibility of this being an empyema. A huge cyst was found within the substances of the right upper lobe which yielded thick pus on aspiration. There were only a few thin adhesions to the apex of the chest. On performing a right upper lobectomy, it was noted that the entire blood supply to the lobe consisted of two small arteries from the pulmonary artery and two small veins which entered the superior vena cava. The right upper lobe bronchus was entirely normal. There was a thin bronchial stalk from the cyst, about 2 to 3 mm. in diameter, which communicated with the right upper lobe bronchus where it originated from the right main bronchus. Following lobeectomy the lower lobes, which were completely free of disease and to which there were no adhesions, expanded and filled the entire chest, as was seen in the postoperative film. Recovery was uneventful and the patient's general condition consistently improved.

Pathologic examination showed the cyst to be lined with bronchial epithelium and the surrounding pulmonary tissue in the remainder of the right upper lobe to be infiltrated completely with inflammatory tissue. We feel that the presence of a normal right upper lobe bronchus and the stalk to this cyst plus its unusual blood supply puts this lesion into the class of a true congenital bronchiogenic intrapulmonary cyst.

DR. ETHAN FLAGG BUTLER, Elmira, N. Y.—In bygone days, in operating upon pulmonary lesions that could be considered congenital in origin, I have five times en-

countered anomalous arteries arising from the aorta and entering lung tissue. I would like to ask Dr. Ramsay, and any other discussors if, in similar congenital pulmonary lesions, they have also noted vascular anomalies.

DR. MARK M. RAVITCH, New York.—Dr. Ramsay has demonstrated to the Association these obstructive changes in the bronchi. I am not confident that the demonstration is equally clear that these are congenital. The one patient who had tuberculosis brought to mind the fairly frequent development in children of fairly advanced bronchiectasis on which operation is ultimately undertaken, sometimes at an early age, with relatively little evidence that the obstruction is actually caused by the tuberculous process, unless one has watched the development over years with serial roentgenograms. I would like very much to hear also what the histologic changes were at the point of obstruction, and the histologic character of the lining of these cysts and bronchi.

DR. CAMERON HAIGHT, Ann Arbor.—It has been our opportunity in the last six months to see two cases of congenital bronchiogenic cysts, in which the lesions have had a solid appearance resembling a tumor. In each instance the lesion was relatively small, not over the size shown in Dr. Harter's case, and had a sharply circumscribed wall by roentgen examination. A preoperative diagnosis of probable indeterminate type of tumor was made in both cases. At operation they were found to be bronchiogenic cysts, completely filled with sterile purulent fluid and having no bronchial communication. One of them was situated in a sequestrated portion of the lower lobe into which an anomalous artery entered from the aorta.

DR. RAMSAY (Closing).—I would like to thank all the discussors and will answer their questions as best I can. With reference to Dr. Butler's question about anomalous vessels, I would like to say that we have indeed encountered several cases of bronchiogenic cyst where the blood supply was anomalous. We feel that this is another type of bronchiogenic cyst, the so-called sequestration cyst. We believe the embryology, or the anomaly, is a different type entirely. That is not a part of the paper but is a part of the subject.

I cannot answer Dr. Ravitch's question too well. In two of these cases the site of obstruction had to be left physically within the patient. In the first case presented, the mucocele was thinned and translucent, and it was almost possible to see through it; it was perfectly flush with the superior segmental orifice. It was so thin that we felt it necessary to close the proximal site with several silk sutures. In the second case the obstruction was narrowed and fibrous, it did not require closure with sutures, but it was immediately upon the upper lobe bronchus and could not be removed for study. So it was not until the third case that we had finally something from which we could make a photomicrograph. We asked the pathologist to photograph it; he sectioned it, and passed the buck to someone, who photographed the cyst but not the septum. We went back to get the specimen and they had discarded all but a small bit of the wall.

If this were a rare condition I would be more unhappy than I am because it might never be possible to demonstrate whether or not these were congenital, or just what the pathology was. However, bronchiogenic cysts are very common, and I believe if each is examined by dissecting the bronchi to the distal side, other septa will be discovered frequently, by many different people. We have a patient now on whom we hope to operate within a month, and we hope we can obtain a photomicrograph in time for publication.

In Case 1, the mucocele was so thin and glistening that I do believe it was congenital. Our concept is, however, that proximal bronchial obstruction, whether congenital or acquired, may produce the entire sequence of changes.

With regard to Dr. Haight's interesting case, which parallels our case of mucocele considerably, there was no bronchial communication. I would like to know if the bronchi were examined carefully, proximal to distal, and if there was an intact septum in that case.

AORTIC COMMISSUROTOMY

HENRY BYRON LARZELERE, M.D.,* TOLEDO, OHIO, AND
CHARLES PHILAMORE BAILEY, M.D.,** PHILADELPHIA, PA.

CHRONIC rheumatic valvular heart disease, the end result of previous rheumatic pancarditis, is one of the most devastating disease conditions found in man. The eventual success in recent years of the cardiologist, the anesthetist, and the thoracic surgeon in eliminating and diminishing many of its sequelae has encouraged even more and continued endeavor.

The mitral valve, particularly stenosis of this valve, has in good measure been successfully attacked by instrumental and digital commissurotomy. Even further advances will be made in the future.

Disease of the aortic valve has to a great extent eluded similar efforts. Transaortic, including retrograde transcarotid, approaches have met with significant failure, resulting in false passage and inconclusive results. Previous attempts via the left ventricle have resulted in some catastrophes due to unguided and blind techniques. In other instances, deliberate creation of aortic insufficiency by surgical means has experimentally and clinically been shown to yield poor and often fatal results.[4, 17, 36]

The concept of commissurotomy, as applied to the aortic valve, seems to include every pertinent physiologic and pathologic factor in correct proportion. It would seem that rheumatic fusion of the three commissures in this valve must yield to suitable instrumentation, resulting in partial or complete separating of the fused commissures. There results a restoration of valve function to as near a normal state as the associated valve pathology can permit.

Just as mitral stenosis can be corrected by instrumental and/or digital means, so a stenosed aortic orifice can be opened. In fact, the presence of three cusps in the aortic valve would mechanically provide even more assurance that an unfavorable insufficiency would not result from commissurotomy. This has been shown through some previous successful efforts and techniques of aortic valve dilatation with an earlier model of the aortic dilator as previously reported.[4, 5]

Cutler and Beck[16] suggested the possible use of a dilator in the treatment of valvular stenosis, referring to the Kohlmann urologic dilator or urethratome.

However, blunt dilatation, combined with blind and, on occasion, false passage, threatened to relegate the procedure, called aortic valve dilatation, to a questionable place on our list of armamentarium.[5] The means by which the commissures are parted and the avenue of approach to the valve have been immensely improved upon since then.

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The present mode of attack and the instrument which will accomplish safe and adequate commissurotomy now assure nearly routinely good results. A triradiate parallel expanding mechanism, containing three specially designed flanged blades, is used to part the fused aortic commissures (Fig. 1). A transventricular approach is used, the instrument being passed into place over a previously inserted beaded guidewire.

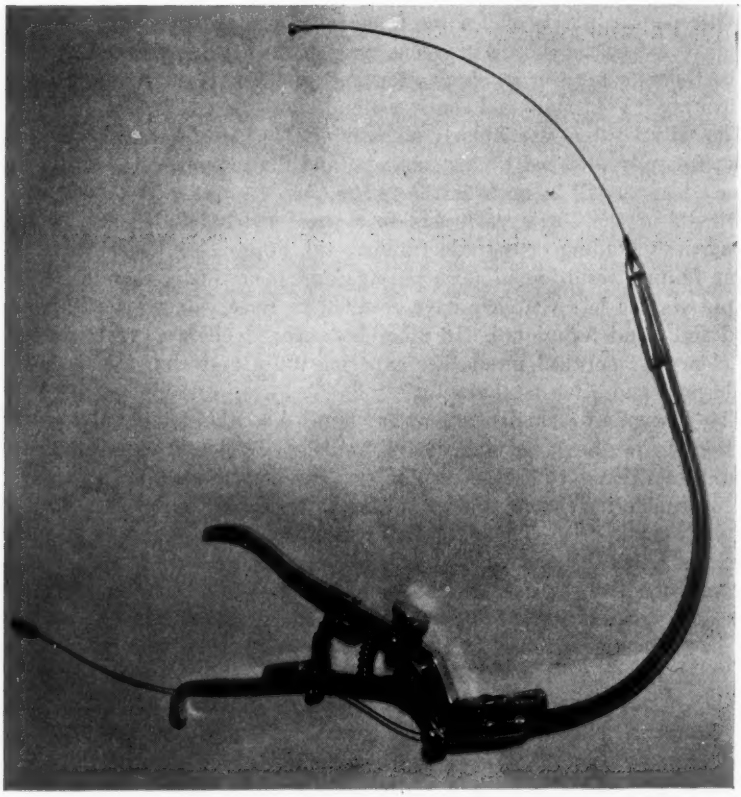


Fig. 1A.—The Bailey aortic "dilator."

ETIOLOGY AND PATHOLOGY OF AORTIC STENOSIS

Although our prime concern is with the adult acquired rheumatic type of aortic stenosis, sufficient numbers of case reports in the medical literature, plus our own clinical experience, have warranted due concern over the differentiation of the several varieties of aortic stenosis. These include the degenerative and congenital types, as well as the rheumatic.

Congenital lesions of the aorta and the aortic orifice, including the outflow tract of the left ventricle, are most aptly presented by Taussig.[39] Pertinent to

this presentation are the following: (a) aortic atresia and marked hypoplasia of the aortic orifice, (b) the bicuspid aortic valve with or without associated aortic regurgitation, (c) congenital aortic valvular stenosis, (d) subaortic stenosis, (e) coarctation of the aorta combined with subaortic stenosis and aortic insufficiency, and (f) supernumerary aortic cusps with fusion.[34]

The presence of aortic atresia and hypoplasia of the aortic orifice is very rare, and the associated nearly constant lesion of defective left ventricular development make any opportunity for surgical correction virtually impossible. Furthermore, there is also usually an underdevelopment of the mitral valve.

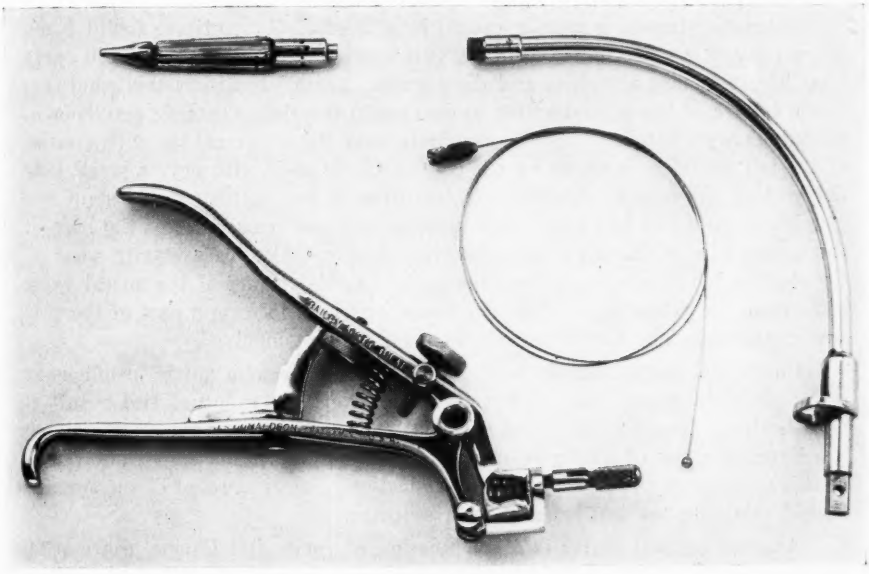


Fig. 1*B*.—The Bailey aortic "dilator."

Such infants rarely survive more than a few days or weeks; others sustain intrauterine deaths. This clinic has observed one case where the left ventricle supplied only the coronary arteries, with a hypoplastic strand of aortic tissue extending from just beyond the coronary ostia out to a large patent ductus arteriosus.

The bicuspid aortic valve of infants and children presents a fusion of the cusps, but the leaflet is thin and is almost always associated with some other malformation of the heart.[39] The adult bicuspid aortic valve may represent inflammatory commissural fusion in a valve of originally otherwise normal structure. Gross[21] believed that the majority of bicuspid aortic valves represented previous inflammatory disease, in some cases occurring early enough to stunt growth.

Abbott[1] and Lewis and Grant[31] recognized the susceptibility of the bicuspid aortic valve to subacute bacterial endocarditis. Also, they[31] noted that the cusps

are usually of unequal size, and that sometimes there is a ridge or raphe which divides the larger of the two cusps. This might suggest an acquired lesion in some cases.

In aortic valvular stenosis[39] there is sometimes found such an unusual size and position of the three aortic cusps as to indicate that the condition must represent a congenital malformation. This is compatible with life for many years and is difficult to differentiate from acquired disease. Only the age of the patient and the known duration of the signs and symptoms point toward the correct etiology. This report includes two cases of clinically diagnosed congenital aortic valvular stenosis treated surgically.

Subaortic stenosis is usually caused by a band of fibrous tissue found 1 cm. below the aortic valve in the adult.[39] This band or membrane, present in early fetal life, normally atrophies and disappears. Keith[26] contrasts this condition to the failure of the infundibulum to develop in the right ventricle resulting in subpulmonary stenosis. That the membrane may stretch across the entire outlet of the left ventricle is shown by the case of Christian,[12] with only a small hole perforating the center. Usually the condition is less extreme.[39] Most of the membrane atrophies and only a few strands of tissue remain across the base of the aorta; this is called partial subaortic stenosis. Also, occasionally some of the chordae tendinae arising from the septal (aortic) leaflet of the mitral valve come from the subaortic membranous tissue, and help to form a part of the subaortic stenosis. The mitral valve cusp itself may be involved.

Congenital aortic stenosis is often associated later with aortic insufficiency and ordinarily causes little incapacity. However, the eventual end result is usually death from superimposed bacterial endocarditis. A point is made that the sievelike effect of aortic or subaortic stenosis may be an important factor in this subsequent and usually fatal complication. Surgical relief of the stenosis should minimize the incidence of such sequelae.

Another clinical entity is a combination of congenital lesions, coarctation of the aorta combined with subaortic stenosis and aortic insufficiency. Here relief of both prime lesions is quite feasible in our opinion. One patient with this condition was operated on in January, 1953, but with correction only of the coarctation. The degree of aortic insufficiency did not warrant aortic commissurotomy attempts; it was hoped that relief of the coarctation with corresponding relief of the associated hypertension would have a beneficial effect on the degree of aortic valvular insufficiency. However, in this case, no definite benefit in this regard was obtained in the immediate postoperative period. It is hoped that, as a readjustment of pressure manifests itself in the circulation, some improvement will become apparent.

Fusion of the supernumerary cusps in the aorta may cause stenosis.[34] Postulated etiology of such fusion is fetal endocarditis or an arrested development.

As to the etiology of acquired aortic stenosis, two chief concepts persist, often contradicting each other. There is the sclerotic calcific and nodular stenosis, called Mönckeberg's aortic valvular sclerosis,[32] of degenerative or

metabolic origin; and there is the rheumatic or infectious stenosis, first suggested as the cause by Christian.[13] The majority of investigators favor the idea of a rheumatic or infectious etiology, although there is much to support the arteriosclerotic concept.[34] Actually the picture usually supports both ideas, there being numerous transitions between healed rheumatic lesions and the calcified nodular valve.

Reich[34] believes that most are due to arteriosclerotic degenerative change in the valve. He refers to Horan and Barnes,[24] who reported 100 cases and found some degree of coronary sclerosis in each case. The arteriosclerosis usually begins at the inner surface of the valve base as a subendothelial atheromatosis. This may progress to involve the entire valve resulting in a rigid deformity. In one form, each individual valve cusp is completely infiltrated and stiffened by calcium. There is little thickening and no distortion in the shape of the cusp, and the commissures are not fused.[5]

Repeated rheumatic infection results in progressive adhesion of the cusps at their commissures, which eventually causes stenosis of varying degrees. After the passage of considerable time, calcification may be superimposed, just as in rheumatic mitral stenosis. This condition is difficult to differentiate, even at autopsy, from the Mönckeberg type of sclerotic valve.[25] Gregory[20] favors the concept of a rheumatic origin in the vast number of cases. He states that the underlying inflammatory damage in the aorta and aortic valve, regardless of etiology, leads to atherosclerosis.

Hall and Ichioka[22] and Clawson, Bell, and Hartzell[14] all favor the infectious concept. Rheumatic origin is favored because of the frequently observed association of rheumatic lesions in the mitral valve, and frequent microscopic evidence of rheumatic inflammation such as proliferative inflammatory reaction and blood vessels in the valves and Aschoff nodules in the myocardium.[9]

The ascending aorta is nearly always free from internal atherosclerosis in aortic stenosis, suggesting the development of the lesions early in life with resulting protection of the aorta against normal fluctuations of blood pressure.[9] It is, therefore, believed that sclerosis of the aortic valve is secondary to rheumatic infection. Hall and Ichioka[22] note the presence of an "impingement plaque" on the aorta where the jet of blood coming through the narrowed aortic orifice strikes the aorta, resulting in a sclerotic plaque, while the rest of the aorta is free of involvement. We have noted this in one case. In the 13 cases of aortic stenosis unassociated with other valvular disease presented here, the average duration of known murmur was fifteen years. The average age of this group was 36 years at the time of operation. Actual presence of a murmur was no doubt present over a much longer time, but had not been brought to the patient's attention. This evidence would highly favor the concept of rheumatic origin as the cause of aortic stenosis.

When subacute bacterial endocarditis occurs, it usually is superimposed upon rheumatic or arteriosclerotic lesions of the valve or engrafted upon anomalous cusps. The acute form of bacterial endocarditis may produce large vegetations on the valve.[34]

There may be associated aortic insufficiency resulting from curling and retracting of the leaflets following rheumatic endocarditis. This results in a marked hypertrophy of the left ventricle. Bell[9] believes this lesion to be due to syphilis as often as to rheumatic fever. He states that in rare instances, as in hypertension, aortic insufficiency may be due to relative valvular insufficiency.

It is probable that the highly calcified impacted aortic valvular stenosis is least amenable to corrective surgery. Yet, 9 of the calcified aortic valves encountered in the 13 cases reported here, with isolated aortic valve disease, definitely improved, as Table I shows.

With minimal or no calcification, the periphery of the valve cusp can again function as a hinge, once the fused commissures are parted; whereas, in those advanced cases of calcific change, the valve, including its annulus, may be one mass of nodular calcification. Yet, even so, some relief is won by overcoming the stenosis. In 5 cases there has been definite evidence of increased insufficiency following aortic commissurotomy. This includes the two cases of congenital valvular stenosis (P. P. and W. E. L.). All of these patients were significantly improved clinically except one, R. F., who died 28 days postoperatively due to reactivated rheumatic infection, pericarditis, and myocardial failure. In five cases a pre-existing aortic insufficiency was significantly improved, in one case apparently completely abolished.

Patients with congenital aortic stenosis may well present a bicuspid aortic valve. The larger of the two cusps may be lacerated at surgery, resulting in regurgitation. It is suggested that dilatation of the congenital aortic stenosis be accomplished "gingerly" in a step-wise manner, increasing the dilatation gradually with repeated check for evidence of developing insufficiency. It may be necessary to repeatedly withdraw the instrument from the valve orifice during these checks, leaving the guidewire in place.

An alternative method is dependent upon the use of a dilator head embracing two, rather than three, parallel bars.[29] This might be safer in cases of evident congenital aortic stenosis, since the separating effect would be applied at the two extremities of the congenital valve slit, rather than at any point against the continuity of a cusp. Such an accessory dilating head has been produced and described.[29]

Sophian,[37] in 500 consecutive autopsies, found 31 cases of fibrocalcific aortic valvular deformity. The total number of cases of valvular heart disease was 57 (excluding luetic aortitis). By histopathologic evaluation he classified 17 of the 31 aortic cases as being caused by rheumatic fever (there was a positive history in 13 cases and associated mitral stenosis in 10 cases). In 14 of the 31 aortic cases, he found evidence favoring an atherosclerotic or degenerative origin; yet, 2 had a history of rheumatic fever and 2 had associated mitral stenosis. He found a variable fusion of the aortic cusps resulting in a dense triangular opening in a fibrocalcific diaphragm or in a false bicuspid valve.

That the pathology of aortic stenosis had a direct relationship with that of the mitral valve is seen in the high incidence of simultaneous involvement. Clawson, Bell, and Hartzell,[14] in a review of 130 cases of chronic endocarditis, noted that more than one-half of the cases showing mitral valve involvement

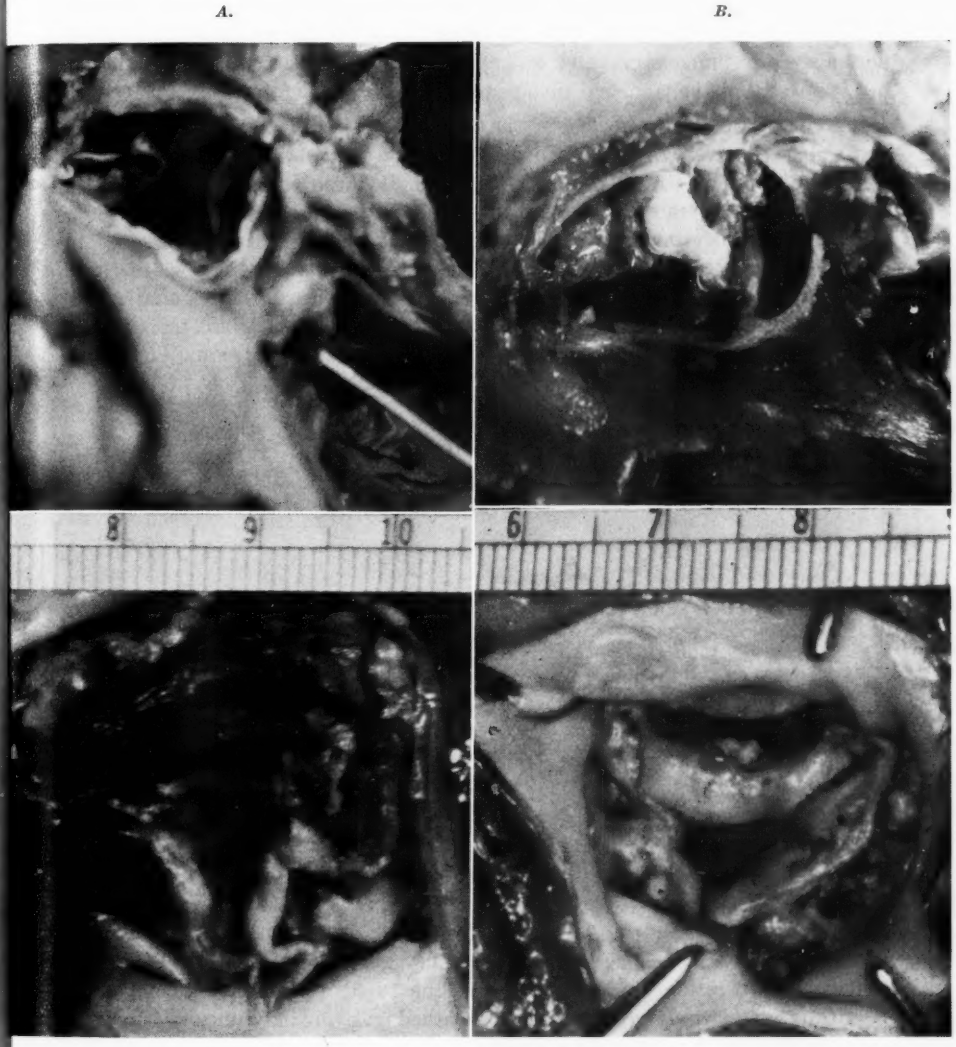


Fig. 2.—Photographs showing the results of surgery on stenosed aortic valves; five cases are shown. *A*, Rheumatic aortic stenosis with commissures all parted by the dilator. Pointer is in line with the posterior pulmonary valve commissure, demonstrating its alignment with the anterior aortic commissure. This "line" passes through the center of each valve ring, and represents line or plane the instrument lies on when used operatively. Case L. K., age 42. *B*, Rheumatic-sclerotic aortic stenosis with the anterior and left posterior commissures well-parted. Gauze has been stuffed into the valve orifice for contrast. Case H. W., age 41. *C*, Rheumatic-sclerotic aortic stenosis with the anterior and left posterior commissures well-parted. Case D. P. R., age 49. *D*, Rheumatic aortic stenosis with the two posterior commissures well-parted; the anterior commissure was open, but rather rigid. Case E. Sl., age 51.

also had aortic valve changes. It is interesting to note that they found 13 cases in this series with tricuspid valve pathology and 3 with pulmonary valve abnormality, the pulmonary valve lesion being unassociated with other valvular involvement, but the tricuspid involved in each instance with other valves.

Autopsy was obtained in 6 of the 7 early and late deaths in patients who had had aortic surgery. In each instance an apparently satisfactory opening had been accomplished. In 2 of these patients, all three commissures had been parted; 4 cases revealed separation of only two commissures. It was evident that appreciable restoration of valve function had been obtained in these. In the remaining cases, a very heavily calcified valve had been split or fractured along two commissures, but the rigidity of the structures suggested that no appreciable increase of valve function had been established (Fig. 2).

Fig. 2*E*.—Rheumatic-sclerotic aortic stenosis with the anterior and left posterior commissures well-parted. The right posterior commissure is incompletely parted. Case J. M., age 37.

The clinical evidence in the surviving cases and this autopsy evidence seem to warrant the conclusion that this instrument, used as directed, routinely accomplishes separation of one or more of the fused aortic valve commissures. Regurgitation is rarely produced by this method and valvular function is usually re-established to a considerable degree.

There are two clinical types of cases which seem to deserve special consideration: (a) the extreme massive nodular calcific aortic stenosis, and (b) the congenital valvular aortic stenosis. In the former, accurate commissurotomy seems to be possible, with little real danger. However, improvement may be slight. Fortunately, relatively few of these fail to show significant improvement with commissurotomy. In the latter, clinically significant aortic insufficiency can readily be produced. Yet, the limited number of such cases in this series precludes dogmatic or tenacious conclusions.

CONCEPTS

Early experimental and clinical experiences with the surgical attack on the aortic valve, embracing cutting or cutting out portions of the valve cusps, convinced us that they offered no feasibility of successful clinical application. A subsequent observation by one of us (C. P. B.) that the fused aortic commissures in rheumatic aortic stenosis tended to separate anatomically with the application of any blunt dilating force within the valve orifice led to the concept of instrumental commissurotomy as presented here.[4, 5]

To implement this principle, a mechanism with three parallel dilating, or separating, bars on a swivel mechanism, which could automatically adjust to the remnants of the fused commissures, was subsequently developed. Such an instrument would apply dilating or disruptive pressure at the appropriate anatomic points (Fig. 8).

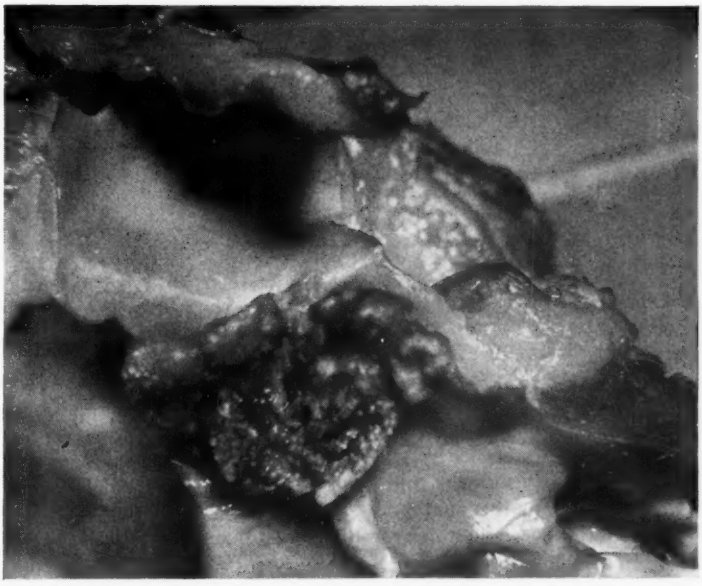


Fig. 2*F*.—Case J. M., with aortic ring opened to show calcareous deposit in the left posterior commissure. Note calcific spur on septal leaf of mitral valve and thickening of chordae tendinae.

An addition to these ideas was the use of flanged bars, wedgelike on cross section, which would tend more definitely to part or separate the fused leaflets.

Efforts to devise a suitable surgical instrument capable of accomplishing the desired effect and a surgical technique which would render it reasonably safe inevitably followed. Various instruments, including spring and "umbrella" type dilators, were tried without complete satisfaction. Different surgical approaches and techniques were tried, including the retrograde approach through the common carotid artery and through the aorta itself. Insertion of the finger

and/or instrument into the aortic valve via the left auricular appendage and mitral valve orifice was attempted and abandoned.

Finally, through the efforts of Major John Shearman Donaldson of Chatham, N. J., an instrument capable of accomplishing these objectives by a transventricular approach was developed and perfected in March, 1952. At the suggestion of one of us (H. B. L.), a beaded guidewire, capable of performing a function comparable to that of a filiform bougie used in urologic procedures or to that of a transesophageal guiding thread used in endoscopic dilatation, was incorporated in the instrument. This provided accurate localization of the aortic orifice from below and precluded the possibility of producing a false passage during attempts to introduce the dilator into the aortic orifice (Fig. 3).

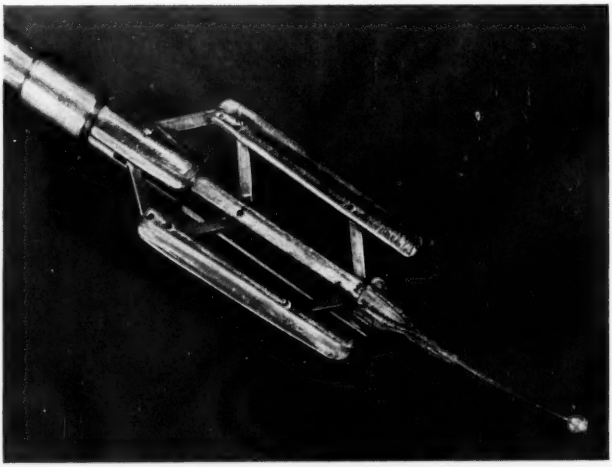


Fig. 3.—Expanding blades and guidewire of the "dilator."

This principle of guidance, plus the automatic adjustment and application of the flanged bars to the anatomic remnants of the commissures, took away much of the "blindness" of the procedure. To one of us (H. B. L.) came the concept of "lining up" the aortic commissures. An understanding of the developmental anatomy of the great vessels at the base of the heart gives the surgeon additional assurance that the adult anatomic landmarks will be recognized.

The aortic valve ring lies slightly posterior to, and to the right of, the pulmonary valve ring; the two rings are directly adjacent to one another. During development, the aorta and the pulmonary artery arise from one common vessel, the truncus arteriosus. A longitudinal indentation, from each side simultaneously, results in the formation of the two vessels, as seen at birth. The original vessel contained four valves or leaflets; the newly formed vessels each contain three valve cusps or leaflets, because of the division of each of the two original lateral leaflets[3, 19] (Fig. 7).

The cusps, resulting from the division of the truncus arteriosus into these two major vessels, are so placed that the anterior commissure of the aortic valve is immediately adjacent to, and in line with, the center point or axis of the two valve rings (Fig. 7). By lining up the pulmonary and aortic valve rings, the commissures of both rings are accurately envisioned. It is the anterior aortic commissure, placed directly adjacent to and behind the pulmonary valve ring, and lying midway between the right and left coronary artery orifices, that is parted by the inner blade of the expanding mechanism. In other words, when the handle is placed in line with the aortic and pulmonary valve rings, one of the expanding blades, that innermost and nearest to the handle, is in line with the anterior aortic commissure. When this situation is established, the other two expanding blades will be accurately applied to the two posterolateral commissures.

Because there is a 360 degree swivel mechanism at the collar or neck of the instrument, the head of the mechanism, at the initial engagement into the valve orifice, automatically tends to become correctly aligned to any remnants of the diseased commissures. In this way simultaneous accurate pressure and parting force is applied directly at each aortic commissure (Fig. 8). Nevertheless, there is a distinct advantage to inserting the instrument with the inner blade on a line with the commissures as described, since it insures this commissural apposition.

Several factors determine in which position the patient is to be placed while undergoing the operation. If the pulse pressure is very low and the blood pressure is at 80 or below after induction of anesthesia, the supine position is best in a case of isolated aortic stenosis. A 10 or more point fall in pressure is often seen when a patient, anesthetized in the supine position, is placed in the lateral position. Also, if the patient clinically and by electrocardiogram shows evidence of severe myocardial strain, or is aged and feeble, the supine position is favored.

However, from the standpoint of technique and for the best operative performance, the lateral position is the position of choice. Exploration and operative correction of associated mitral valve disease can be accomplished far more readily. The isolation and control of the great vessels arising from the aortic arch are almost impossible in the supine position; these vessels are the avenues for any emboli to reach the brain during the procedure. They may be temporarily and intermittently compressed at strategic moments during the operation to prevent brain embolization.

Amputation of a thrombosed left auricular appendage is rather difficult in the supine position. Commissurorrhaphy for mitral insufficiency can scarcely be performed, except with the patient in the lateral position.

Because of the high incidence of coexistent mitral disease in patients with aortic involvement (over 50 per cent) and because this lesion may be overlooked in the preoperative evaluation of the patient, it is our belief that the mitral valve should be explored digitally at the time the aortic commissurotomy is performed. This premise would seem largely to preclude the use of the supine position and the anterior approach, except under unusual circumstances as mentioned.

Generally unsatisfactory experience in patients with such multivalvular lesions, when either is left uncorrected, whether by accident (overlooked) or by design, has convinced us that whenever present such bivalvular lesions should be simultaneously attacked. Not only do such patients fail to show satisfactory improvement when one valve is left uncorrected, they frequently fail to survive operation. Two cases are cited where relief of mitral stenosis has merely accentuated a coexisting aortic stenosis, since the latter was not relieved. The patients subsequently died in failure. They were J. I. R. and R. I., death coming twenty-four hours and seven weeks postoperatively, respectively. Also, I. M. B. of this series, with extensive old tuberculous pleuritis and fibrotic lung lesions, had both aortic and mitral stenosis. Exposure for aortic commissurotomy was feasible, but not for the mitral valve correction. There has been an improved pulse pressure, but there remain the unmistakable signs of mitral stenosis; this lesion was originally felt to be of only moderate severity. Further reference to these cases will be made.

Evidently a severe obstruction of a given degree at any one point to the flow of blood in the heart is about as much of a handicap to the circulation as a whole as a similar degree of obstruction at two or more points; correction of any one such lesion alone is, therefore, both dangerous and clinically ineffective.

A frustrating observation is the occasional recognition, by auscultatory and other clinical evidences, of an advanced aortic stenotic lesion which was not noted or fully appreciated prior to the surgical correction of a severe mitral stenosis. It is postulated that the diminutive ventricular output per beat in advanced mitral stenosis is not significantly impeded by a moderate to severe aortic stenosis. Therefore, the auscultatory and other evidences of the latter lesion do not become apparent until the improved left ventricular filling, subsequent to relief of the mitral obstruction, results in a relatively more severe degree of aortic obstruction.

It is interesting to note that the patient with mitral stenosis reported by Pribram[33] died on the sixth postoperative day of both fresh endocarditis and pneumonia, after relief of a stenosed mitral valve resulting in mitral regurgitation. It is possible that if simultaneous relief of the aortic stenosis, which was present, had been accomplished, sufficient ventricular output might have reduced the tendency for pulmonary congestion and the development of endocarditis. The aortic stenosis was first discovered at autopsy.

While many of these patients with aortic stenosis first discovered after mitral commissurotomy seem to do well clinically at first, it is our fear that sooner or later progressive symptoms of aortic stenosis will require secondary surgery. For this reason we recommend that the intrapericardial portion of the aorta be routinely carefully palpated at the time of surgery in all patients subjected to mitral valvular surgery. A systolic thrill in the root of the aorta thus found unexpectedly might well be considered as evidence of aortic stenosis which can be immediately subjected to instrumental correction.

Simultaneous attack on both aortic and mitral stenosis, in a given case, is recommended. In the 42 instances in which aortic commissurotomy was per-

formed by the technique described in this article, 23 mitral commissurotomies were also done, each at the same operation. In these operations, the mitral valve was usually corrected first. In some instances following adequate commissurotomy for the mitral stenosis, a definite insufficiency was noted. However, as soon as the pressure built up in the left ventricle was relieved by aortic commissurotomy, the mitral insufficiency immediately disappeared. The apparent safety of simultaneous correction now makes such repair mandatory.

In the face of clinically significant, combined valvular disease, such as aortic and mitral stenosis, there comes the very important problem of which valve to correct first. It is agreed that there is seldom an occasion to stage the operations on these two valves; but, the question is, "Just which valve is to be opened first in the one stage?"

There is much to support relieving the aortic stenosis first. The aortic procedure is comparatively simple in both timing and technique. It is good to have the aortic valve open early in the consideration of the possible development of hypotension, cardiac arrest, or ventricular fibrillation. There seems to be relatively slight danger of producing a significant aortic insufficiency, which might not be tolerated with an obstructed mitral orifice. In no case reaching the autopsy table has there been seen a false commissural opening; other than in one of the cases of congenital valvular aortic stenosis, there was no patient who presented postoperatively a diastolic pressure below 60 mm. Hg. It is felt that the myocardium is better protected, because the aortic valve commissurotomy will immediately allow better coronary filling.

However, there is also much evidence to support opening the mitral valve first. In all but two of the combined cases herein presented (C. W., M. Gi.), the mitral valve was attacked first. The following discussion will elaborate on these factors for decision.

Mitral commissurotomy is a more involved and time-consuming procedure than aortic commissurotomy, but it is less severe on the heart as a whole. Entry into the heart via the left auricle is extremely well tolerated, even by the so-called irritable heart.

Aortic commissurotomy, although a technically more simple procedure, is beset with severe momentary stress effects to the myocardium. The relative incidence of ventricular fibrillation and cardiac arrest, and/or other abnormality associated with hypotension, is much higher in aortic than in mitral commissurotomy. When such an episode occurs, the surgeon must proceed deliberately to open the aortic valve, even if some seconds are lost before applying the cardiac defibrillator or stimulants. Cardiac massage "between the acts" must be performed. If the mitral valve has been left as the second point of attack, ventricular filling and cardiac output are still limited by the degree of mitral stenosis, as yet unrelieved. This is true whether the aortic valve has been opened or not. Such a situation might well guarantee a fatal outcome.

Furthermore, should an insufficiency of the aortic valve be produced in the unusual instance, presence of unrelieved mitral stenosis would tend to prevent compensatory left ventricular dilation. Whereas, with the mitral valve

opened first, such an insufficiency would presumably be better tolerated because of better ventricular filling.

Also, should passage of the guidewire prove to be difficult, simultaneous insertion of the index finger into the left atrium via the auricular appendage would help to guide the wire into the outflow tract of the left ventricle. Palpation of the aortic (septal) leaflet of the mitral valve and insertion of the finger into the left ventricle through the mitral orifice would allow such guidance.

Therefore, we prefer in general to do the mitral commissurotomy first in the face of combined valvular lesions, especially if both valves are stenosed. In the face of mitral regurgitation, and following exploration of the valve, it is best to open the aortic valve before attempting the mitral commissurorrhaphy (saphenous vein or pericardial strip graft suturing of the incompetent mitral commissures) as presented elsewhere.[8]

However, in the face of severe aortic stenosis and severe mitral insufficiency, the aortic valve should be attacked first to rule out functional mitral regurgitation due to aortic obstruction.

Combined aortic stenosis and aortic insufficiency has been benefited in good measure by aortic commissurotomy, with a readjustment of the insufficiency as return of the valve function is allowed. This has been noted in 4 cases out of 17 cases when aortic insufficiency existed preoperatively. In 3 others, there were increases in the insufficiency; one was significant. The other 10 patients remained unchanged in this regard. Such return of function is often seen in the mitral valve when relief of coexistent stenosis is accomplished, the regurgitant flow being markedly reduced by better coaptation of the mobilized valve leaflets. In this instance a bivalve return or competency is brought about, the large septal leaflet having by far the greater degree of possible readjustment. In the aortic valve a near equality in size of the three cusps gives even greater possibility of increased competency once the commissural fusion is relieved.

Although it is conceivable that a refusion of valve commissures is possible, as yet, no known case of such refusion has been reported to our knowledge, either in aortic or mitral surgery. That reoperation, on cases with previously unsuccessful or inadequate valve repair, is possible can be illustrated in a few instances where a mitral valve was opened to a degree allowing excellent recovery over a previous postoperative state labeled unimproved. Also, in one of the cases reported here there was a redilatation of the aortic valve, using the newer instrument; the valve had been "dilated" a year and one-half previously with an earlier, more crude model. In each instance of reoperation, success has been our experience.

THE INSTRUMENT

Four powerful main assemblies compose this instrument, each made of stainless steel.* They include a beaded guidewire, a triradiate dilator mechanism, the handle, and the shaft of the dilator (Figs. 1 and 3).

Although called a "dilator," the instrument performs actual valve commissurotomy by parting the commissures accurately, thus sparing any untoward tearing or stretching.

*Designed, engineered, and manufactured by Major John Shearman Donaldson of Chatham, N. J. Obtainable from Major Donaldson and from George P. Pilling & Son, Co., Philadelphia, Pa.

The triradiate dilator mechanism consists of the head and a collar. Included as parts of the head are the nose, the hollow center shaft, and the dilating or expanding blades. The mechanism, similar in some respects to the pulmonary biradiate dilator of Brock, has connecting levers which thrust the blades outward.

The tapered and flanged nose fits onto the center shaft which, in turn, slides through the collar. Parallel to and surrounding the center shaft, lie the three triradiate flanged blades, which have a wedge shape on cross section. The longitudinal flange of each blade has a dull fin as the leading edge.

The tube-shaft assembly consists of a rigid curved outer tube with an inner flexible spring shaft. This inner shaft is the "working link" between the dilator head and the handle of the instrument. Although flexible, an inner strut makes the spring shaft neither compressible nor expandable.

An adjustment screw on the handle can be set so as to limit the degree of the dilator expansion at any point, the maximum being 1⅜ inches. A safety screw, when set, prohibits expansion completely, keeping the dilator head collapsed and closed. Although a handle spring keeps the dilator closed upon release of the handle, the safety screw guarantees closure. On passing the instrument either into or out of the ventricle, it must be closed to prevent major myocardial laceration. The same is true when the valve orifice is engaged and upon removal of the dilator head from the valve ring. If desired, this "safety" may be removed permanently from the instrument.

Traversing the length of the instrument, and passing beyond the tip of the nose, lies the beaded guidewire, along which the entire instrument can be easily slid.

TECHNIQUE OF AORTIC COMMISSUROTOMY

Anesthesia in aortic commissurotomy has been very similar to that used in the mitral cases.[27] Intravenous pentobarbital sodium and hexamethonium bromide (C_{10}), along with a separate continuous slow administration of 0.2 per cent procaine via venous canulization, make up the intravenous drugs used. Endotracheal intubation with nitrous oxide and oxygen under positive pressure controls respiration.

This type of anesthesia[28] is preferred because it allows rapid induction and endotracheal intubation, lessening the chances of cardiac arrest and ventricular fibrillation. It permits an immediate return to consciousness postanesthetically which makes possible early recognition of any catastrophic embolization peripherally or centrally. The proper treatment for these emergencies can then be instituted immediately.[6, 40] There is less nausea and vomiting postoperatively with this anesthetic combination. A rapid entry into the chest is possible because it allows the use of electrocoagulation for the control of bleeding points, since it is a nonexplosive combination. Endotracheal anesthesia provides an unobstructed airway and promotes oxygenation in surgery of the open thorax.

Blood transfusion is begun more or less routinely once the operation has proceeded to a stage where the pericardium is opened. We do not await blood deficit in this particular disease condition. Also, immediately available is a set for arterial transfusion, which any member of the team can start at once.

Induction of anesthesia is never begun unless at least one surgeon is fully gloved and gowned, and the operating room nurses, both instrument and circulating, are in the room fully prepared to begin surgery. This is to effect cardiac recovery immediately in the face of an arrest or ventricular fibrillation.[11]

After induction in the supine position, the patient is placed in the lateral position when the mitral valve is to be explored and the left chest is entered via the fifth intercostal space. Occasionally, to better expose the left ventricle, the cartilage and the posterior extremity of the sixth rib are severed.

After retracting the lung inferiorly and posteriorly, the great vessels arising from the aortic arch are identified: The vagus nerve runs lateral and anterior to the left innominate vein, the innominate artery lying deep to these structures. In relation to the trachea, the innominate artery lies anterior and to the right (Fig. 5, *A*).

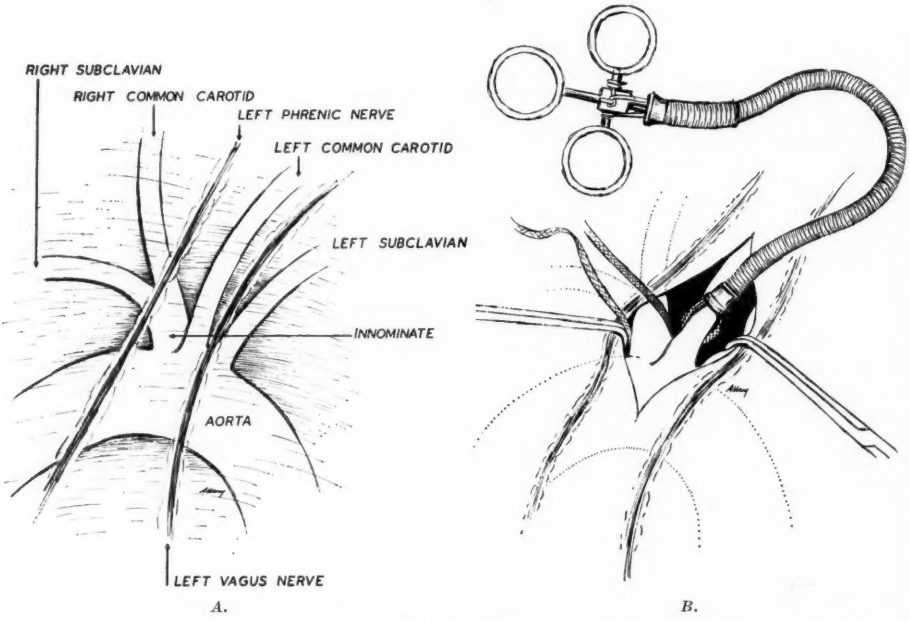


Fig. 4.—*A*, Isolation of the great vessels leading to the brain. *B*, Control of the great vessels leading to the brain. Prevention of cerebral embolization.

Running obliquely across the mediastinum, on a level with the aortic arch, is the left superior intercostal vein. This vein is often severed and ligated to facilitate exposure of the innominate and left common carotid arteries. By using a vertical incision of the pleura, the superior mediastinum is entered just above the aortic arch. Here the vagus nerve and subclavian artery are easily found posteriorly. These structures are retracted and the deeper lying left common carotid artery is freed up by blunt and sharp dissection, care being taken to avoid injury to the deeper lying recurrent laryngeal nerve. Using a right-angled clamp, a narrow umbilical tape is placed around the vessel. The tape is then run through and fixed to a Rumel-Belmont tourniquet[35] (Fig. 5, *B*).

Likewise, by retracting the phrenic nerve and the left innominate vein anteriorly and the left common carotid posteriorly, the innominate artery is

found at a deeper level anterior and deep to the trachea. By blunt and sharp dissection this vessel is encircled with an umbilical tape and another Rumel-Belmont tourniquet is attached. These tapes now permit definite and controlled interruption of the blood flow in the two large arteries supplying blood to the brain. Intermittent control with the prevention of cerebral embolization is possible at any time there are operative or manipulative procedures either on or within the heart.[6]

Specific moments when the cerebral flow is interrupted are: entrance of the guidewire into the aortic valve orifice, engagement and dilatation of the aortic valve, and, in concomitant mitral surgery, when the auricular appendage is manipulated, clamped, or entered. Also, during exploration and commissurotomy of the mitral valve by digital or instrumental means, the vessels are occluded. At no time should they be occluded for periods longer than two minutes, the preference being a time limit of one minute with "rest" periods of two minutes to allow adequate cerebral oxygenation.

The pericardium is now opened approximately 2 cm. anterior to and parallel to the left phrenic nerve, giving wide exposure of the posterolateral surface of the left ventricle and the first portion of the aorta. The aortic ring and aorta are palpated, and the character and size of the ring and the size and force of the systolic jetlike thrill over the aorta are noted. Also, any diastolic thrills of regurgitation are noted. The dilator is then set so that the blades expand to within a millimeter or two of the external aortic ring diameter. This is determined by palpation, as a rough but practical finger-thumb measurement. The lock and safety screws are now set.

Note is now made of the two valve rings at the base of the heart. The aortic and pulmonic rings lie one behind the other. The surgeon is able to accurately determine the angle or plane the instrument will eventually be placed in, when the time comes for the head of the dilator to enter the aortic valve orifice (Figs. 7 and 9).

After thoroughly injecting the area with 1 per cent procaine, a purse-string suture is placed into the wall of the left ventricle on the anterolateral aspect, a relatively avascular area being chosen, approximately 5 cm. away from the apex of the heart (Fig. 5, *A*). This is a tripoint suture, each bite being deep and well spaced to produce a circumscribed area of 3 cm. in diameter. Care is taken to avoid damaging any secondary or tertiary coronary vessels by placing the suture so it passes under any adjacent vessel or avoids it.

Three bites, and no more, are advised in the placement of this purse-string suture. It is dangerous to place the suture in more than three places, for with more than three such bites the force necessary to overcome the friction of the additional suture placements, and yet adequately secure hemostasis, may result in tears through the muscle layers of the myocardium. Severe bleeding is of course the result in such instances. Extreme softness of the ventricular myocardium frequently is seen in cases of aortic stenosis and necessitates special precaution.

Delicate, yet snug and adequate, control of the purse-string suture is very important. The use of the Rumel-Belmont tourniquet has become standard;

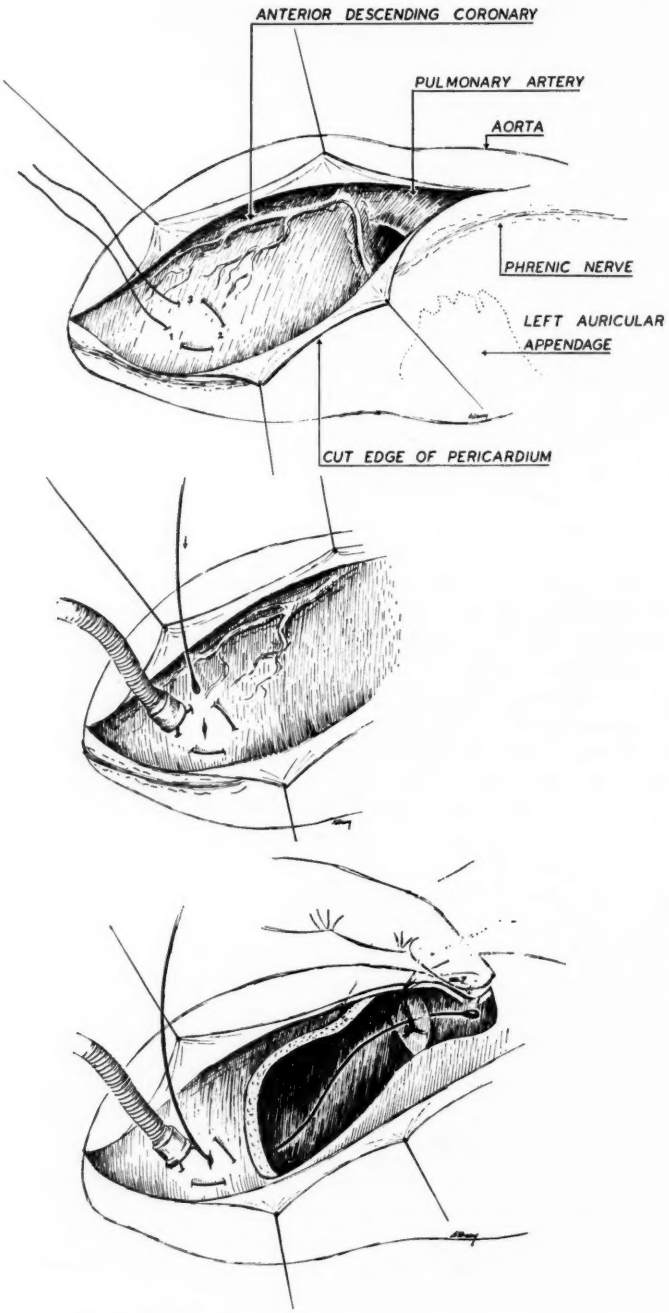


Fig. 5.—Technique. *A*, The purse string. *B*, Insertion of the guidewire. *C*, Passage of the guidewire through the aortic orifice.

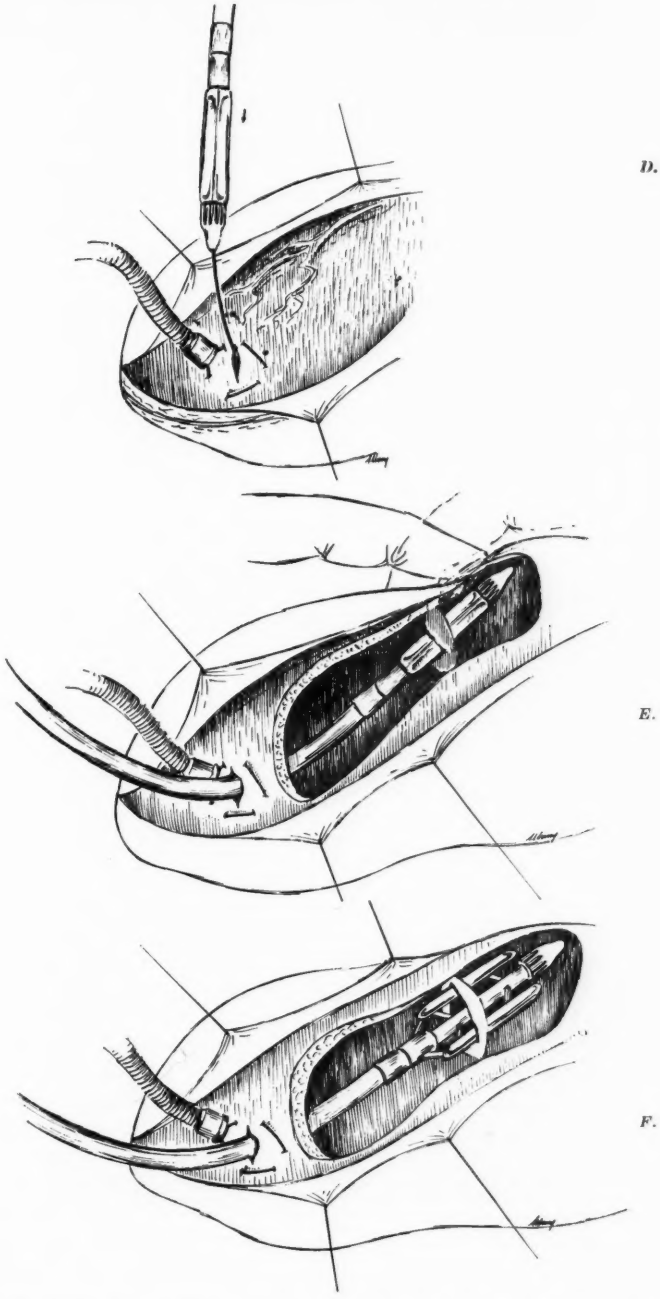


Fig. 5.—*D*, Enlargement of ventricular incision to receive the head of the instrument. *E*, Engagement at the aortic valve. *F*, Effecting aortic commissurotomy.

the suture is passed through the tourniquet and held by an assistant. The suture is of braided silk or cotton, size No. 2, with a relatively large round atraumatic needle. We prefer some blood loss from a less than snug purse-string suture to major hemorrhage from the irregular wounds resulting when a hemostatic suture, very tightly held, tears through the ventricular wall.

Using a No. 11 blade, a tiny longitudinal stab wound is made through the left ventricle at the center of the purse-stringed area. The beaded guidewire is extended so as to project a good six inches from the nose of the dilator. It is gently inserted into the heart via the stab wound and is carefully passed up and through the orifice of the aortic valve (Fig. 5, *B* and *C*). Assurance of the position of this guidewire is had by simply compressing the first portion of the aorta between the left index finger and thumb. Forceful passage of the guidewire is unnecessary and can cause perforation of the heart wall.

Next, the small incision in the ventricle is deliberately enlarged longitudinally by the scalpel to 1 cm. in length through the entire ventricular wall. Reliance on direct instrumental pressure to enter the ventricle is not safe, for such pressure may stimulate ventricular arrhythmias or may cause a stellate tear in the heart wall leading to a major bleeding and to difficult repair (Fig. 5, *D*).

The triradiate dilator head is then passed along the wire, into the left ventricle and on to engage within the orifice of the aortic valve (Fig. 5, *E*). It is during this and subsequent movements of the dilator within the heart that the vessels leading to the brain are temporarily obstructed to prevent any possible cerebral embolization from bits of calcium which may be dislodged from the free edge of the valve.

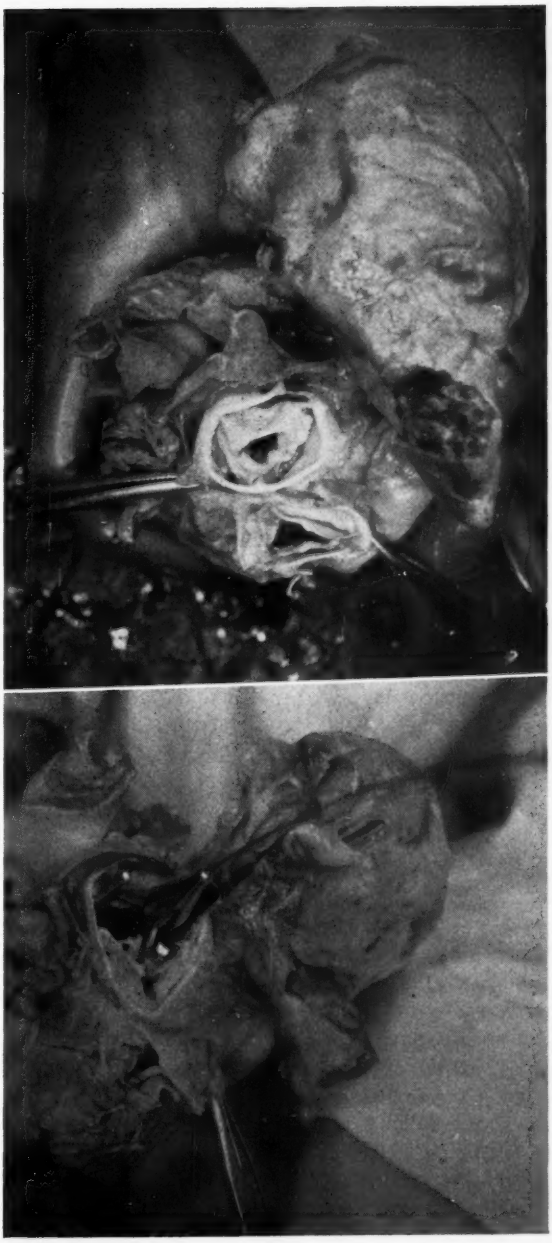
The location of the dilator head is confirmed by external palpation. If necessary, the head can be boldly forced through a tight orifice. On one occasion the amount of valve calcification required a relatively severe continued thrust in order to adequately enter the orifice. The guidewire effectively prevents any false passage or perforation by the dilator.

As the dilator is advanced along the guidewire, the wire is pulled back a similar degree by the assistant to prevent any coiling of the wire in the aorta or any tendency for it to advance up the vessels taking off from the aortic arch. On two occasions the guidewire became momentarily caught in the innominate artery by the umbilical tape constricting that vessel. This caused the wire to coil and become kinked within the aorta, making a difficult valve engagement by the dilator.

While passing the dilator head through the chamber of the left ventricle, the surgeon again quickly checks the alignment of the pulmonary and aortic valve rings, and swings the handle and shaft of the instrument to lie on a plane parallel to this. This insures proper commissural engagement and helps to make certain the correct line of commissurotomy (Fig. 9).

After it is properly engaged, the safety lock is released and the blades of the head are extended (Fig. 5, *F*). The shocklike impact of this opening of the valve is often easily felt through the length of the instrument. On occasion, forceful compression of the handle is required in order to part badly stenosed or calcified commissures. With one hand at the aortic valve ring, the extended

A.



B.

Fig. 6.—Photograph showing aortic stenosis and instrumental relief; nonsurgical specimen. *A*, Rheumatic aortic stenosis (a nonsurgical case). A case of trivalve stenosis aortic, mitral, tricuspid. The patient was a 53-year-old woman. Aortic orifice shown uppermost. Note the adjacent commissures of the aortic and pulmonary valve rings are on line with each other and the center of each ring. *B*, Instrumental relief of rheumatic aortic stenosis (a nonsurgical case). All commissures well-parted.

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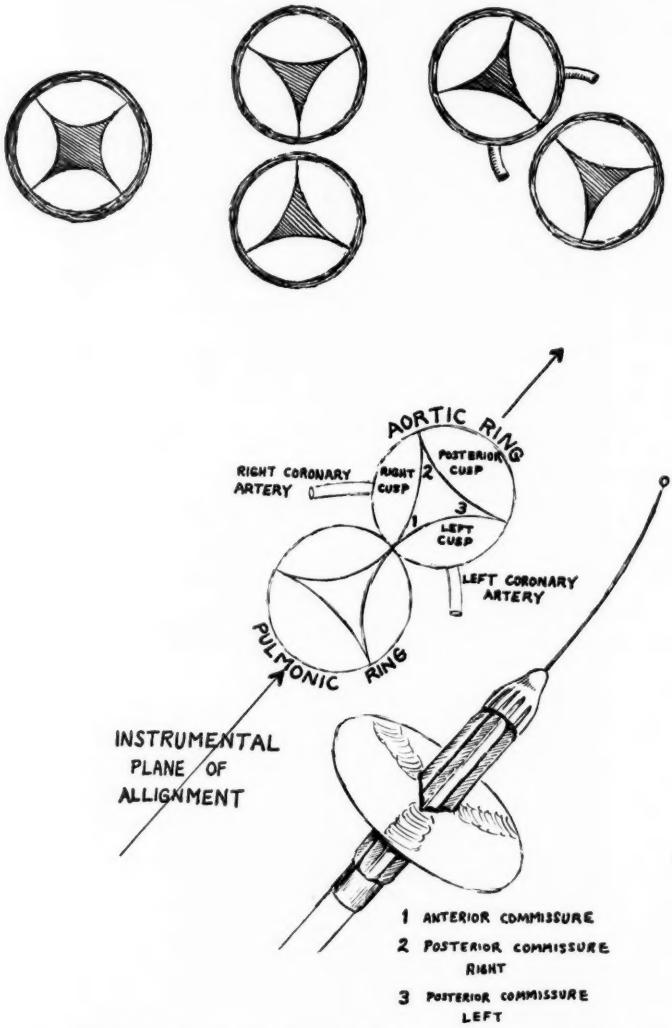

Fig. 7.—Valve rings with the dilator "on-line" with the aortic commissures, embryologic reference.

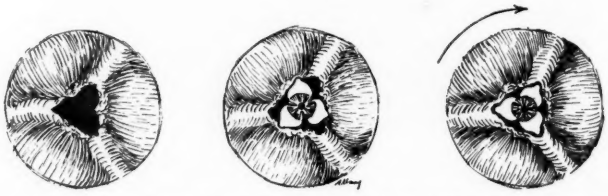

Fig. 8.—Swivel effect at commissural remnants on instrumental engagement.

blades of the instrument can be easily felt. The blades can be extended two or three times, each time pushing forward or pulling back the instrument slightly to better guarantee adequate commissurotomy. The aortic valve lies more deeply within the heart than is ordinarily recognized, providing the possibility of failure to enlarge the valve orifice if the instrument should be extended too far through the valve.

There is an immediate change in the character and force of the thrill over the aorta. It becomes more diffuse and usually less intense at any one point. Also, very often the anesthetist can detect a sudden increase in pulse pressure peripherally.

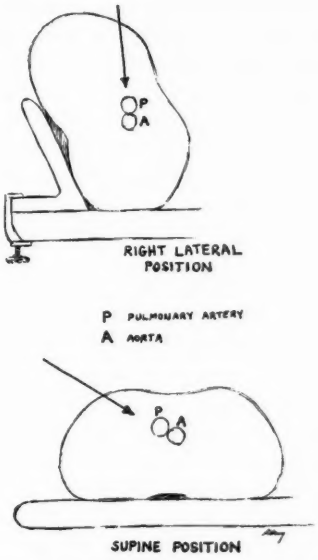

Fig. 9.—Operative position and commissural alignment.

Upon release of the handle, the safety lock is set again and the instrument is withdrawn from the heart. Should this precaution not be taken prior to instrumental withdrawal, it is conceivable that on some occasion one might try to pull out the instrument with the dilator head open. A serious laceration of the ventricular wall could result.

Also, if upon attempting to withdraw the instrument from the heart, it is found to be fixed, the assistant should be asked to release the tourniquets on the neck vessels for the beaded guidewire might be caught by his tourniquet. This has happened to us on two occasions. The instrument, when properly free, comes out of the valve and ventricle very easily.

While the dilator is within the ventricle, and again during the closure of the heart wall incision, the purse-string suture is kept snug by tension on the Rumel-Belmont tourniquet. A word is again passed to the assistant, concerning the greater danger of too tight a purse-string suture than of one held "just snug

enough.'' It is better to have incomplete, yet moderately satisfactory, control of bleeding with a gently held tourniquet, than attempted complete hemostasis by an overly tight purse string, soon followed by laceration of the heart wall and heavy bleeding.

After repair of the heart wound by interrupted or small mattress atraumatic sutures, the purse-string suture is cut and removed. There is left but a small linear wound in the heart wall. In cases presenting excessive softness of the myocardium, it is felt preferable to gently tie a knot in the purse-string suture in order to reinforce the actual suture line.

The pericardium is loosely approximated to allow drainage into the pleural space. Occasionally a sheet of Gelfoam is placed between the pericardium and the operative site on the ventricle. Within seventy-two hours the pericardium is usually sealed shut by fibrinous pericarditis. Should there be a tendency for pericardial fluid to form postoperatively, the surgeon should not rely completely upon drainage into the pleural space, particularly after this immediate seventy-two hour period. Pericardiocentesis was necessary to save a life in one such case. The classical signs of such an effusion are usually absent. However, unexplained enlargement of the heart shadow (by radiography) in association with profuse sweating and a shocklike state required diagnostic pericardiocentesis by the costozyphoid route.

Occasionally passage of the guidewire will be difficult. Curling of the wire and/or calcific deposition on the valve edges, or below the valve, may make for difficult entry into the aortic orifice. Suggestions for overcoming this include the reforming or recurving the guidewire to a slightly different arc in its first portion, and again attempting the passage. Also, the index finger may be inserted into the left atrium via the auricular appendage, and be used to guide the wire into the ventricular outflow tract and subsequently into the aortic orifice. Even the very irritable heart tolerates atrial entry well.

Efforts to palpate the aortic orifice directly, via the left atrium and mitral valve orifice, are very dangerous. This, the Ramirez maneuver, has met with fatal results due to tearing of the septal mitral valve leaflet and chordae tendinae.[4, 10]

However, Tuffier[41] reported a case wherein the aortic orifice was digitally enlarged by invaginating the wall of the aorta into the valve opening. The patient was alive and well some eight years later. We have seen one case where autopsy evidence indicated such an approach might have been used at operation. In general such an approach must be considered far too dangerous.

However, since this writing, one of us (C. P. B.) has successfully used the aortic retrograde approach to the aortic valve in several cases. The aortic wall is grasped by a Beck aortic clamp, and a longitudinal incision is made on the anterior wall of the ascending aortic arch.

By use of a large square of pericardium previously prepared in poncho fashion, or by use of a special rubberized sleeve for both the finger and the dilator, sutured to the incised aorta, the valve can be directly palpated and instrumental commissurotomy effected. Purse-string suturing of the pericardial poncho or compression of the sleeve controls bleeding very well.

Great advantage here lies in the direct ascertainment of valve pathology, and in the lessening of the stress effects on the myocardium which are present in transventricular approaches. Full evaluation of this technique is to be published; at present, it resides in the field of experimental surgery.

PREOPERATIVE EVALUATION AND INDICATIONS FOR AORTIC COMMISSUROTOMY

The indication for aortic commissurotomy is the presence of clinically significant aortic stenosis, certain contraindications withstanding. The condition is found in a far greater number of men than women, a finding just the opposite in isolated mitral valve pathology.

Diagnosis ordinarily rests on the finding of a harsh systolic murmur over the aortic area of the chest, with or without a thrill, and with a diminished or absent aortic second sound. It is not unusual to find evidence of a slight to moderate amount of aortic insufficiency. The systolic thrill and murmur are often transmitted to the neck vessels. Usually, a normal sinus rhythm is present.

Calcification of the aortic valve and its ring is an extremely valuable roentgenographic and fluoroscopic sign associated with aortic stenosis, noted by Contratto and Levine.[15] Sosman and Wosika[38] state that when present it generally indicates an extremely severe grade of aortic stenosis. The consistency of agreement between the roentgenographic diagnosis of calcific valve change with the findings at surgery has been amply demonstrated by Lehman and Curry.[30] All the cases in this series of aortic stenosis, determined to have significant degrees of calcific change preoperatively by our roentgenographic department, were shown at surgery to have a heavily calcified ring or valve. In a few instances it was also demonstrated at the autopsy table. Also, roentgenkymography can beautifully demonstrate diminished aortic pulsation, as reported by Hirsch and Gubner.[23]

There is often a "pulsus tardus" or diminished pulse pressure in severe aortic stenosis. The cardiac output and curve of aortic pressures, as determined by cardiac and brachial artery catheterization, aid greatly in making a good physiologic diagnosis and also, postoperatively, in indicating the degree of correction accomplished.[18] That the blood pressure is influenced to a considerable degree by aortic stenosis was shown by Dry and Willius.[17] They noted that the height of the blood pressure bore a direct relation to the degree of stenosis, although, even if an extreme degree of aortic valve narrowing existed, it was possible to have severe forms of hypertension along with all the associated changes in the peripheral vascular system that occur in cases of hypertension unassociated with disease of the aortic valve.

It was also noted that the amount of cardiac hypertrophy closely paralleled the degree of the aortic stenosis.[17] Atherosclerosis of the aorta and coronary arteries occurs in reverse proportion to the degree of aortic stenosis. Coronary occlusion was rare in the 228 cases of Dry and Willius,[17] but angina pectoris was common and, in turn, depended upon the amount of the stenosis present. They found that the absence of the aortic second sound usually meant severe aortic stenosis. These observations have been all confirmed in our own series.

Roentgenographic and electrocardiographic evidence of heart chamber enlargement and strain aid in the diagnostic evaluation; although, at first, in the sequence of pathologic events in pure aortic stenosis, there is only left ventricular hypertrophy with little chamber enlargement and hence little change in the size of the heart, there is usually evidence of left ventricular strain and hypertrophy in the electrocardiogram. Later, dilatation and rotation of the ventricle result in more obvious signs on the roentgenogram.

The electrocardiogram may show not only left ventricular hypertrophy and strain, but also evidence of myocardial insufficiency, bundle branch block, or even more definite cardiac damage with an infarction picture. That the electrocardiogram is far from infallible is evidenced by the observations of Anderson, Kelsey, and Edwards.[2] They attributed this to the apparent occurrence of prolonged yet reversible myocardial ischemia indistinguishable clinically from myocardial infarction, together with the frequent presence of electrical cardiographic aberrations, such as left bundle block which decreases the value of the electrocardiogram when the diagnosis of myocardial infarction is attempted.[2]

Myocardial failure, as outlined by Christian, is diagnosed by the presence of increased dyspnea on mild exertion, paroxysmal nocturnal dyspnea, episodes of acute pulmonary edema, and hepatomegaly. Of the 13 patients with isolated aortic stenosis included in our report, four were in myocardial failure on admission, whereas six of the 19 patients with combined aortic and mitral stenosis belonged in such a classification. All of the three patients with aortic stenosis and mitral insufficiency were so placed. Table IV.

Contratto and Levine,[15] in a review of 180 cases of aortic stenosis, found that the duration of signs and symptoms prior to death was as follows: palpitation, 8 years; dyspnea, 24 months; edema, 9.3 months; syncope, 9.1 months; congestive râles, 6.3 months; and pitting edema, 4.3 months. Also, in the series of 49 isolated aortic stenosis cases of Anderson, Kelsey, and Edwards,[2] all of whom were correlated by autopsy, as well as clinical findings, myocardial failure occurred in 73.5 per cent, the duration of failure prior to death ranging from seven days to eleven years, the average survival, after onset of a failure, being 2.2 years.

In our series of cases the average duration of failure prior to surgery was 1.2 years for isolated aortic stenosis, 1.1 years for combined aortic and mitral stenosis cases, and 9 months in the aortic stenosis and mitral insufficiency combination. Table IV.

Symptoms which are important include angina pectoris, dizziness, syncope, and convulsions. Myocardial and cerebral circulatory insufficiency may be very prominent. Auricular fibrillation in isolated aortic valve disease is unusual.

Contraindications to aortic commissurotomy are the following:

1. Persistent advanced congestive failure in spite of diligent medical treatment. Failure, which, in spite of adequate digitalization, bed rest, diuretic therapy and restricted salt diet, continues on the same level or becomes fixed or worse, represents a contraindication unless severe calculated risk is considered acceptable. Note Case ESl.

2. A tremendous left ventricle of Grade 4 or 5 proportions. Usually such a ventricle has "worn out" its ability to hypertrophy and is represented only as a degenerated soft myocardium.

3. Gallop rhythm; there is little to offer, because of the advanced myocardial deterioration.

4. Advanced and serious myocardial insufficiency may warrant postponement.

5. Active rheumatic fever episode.

6. Subacute bacterial endocarditis.

7. Age, beyond 50, represents definite and significant added risks.

RESULTS IN ISOLATED AORTIC DISEASE

Aortic commissurotomy was performed in 13 cases with isolated aortic valvular disease. Of this number, 7 had a diagnosis of "pure" stenosis, as determined both clinically and at the time of surgery. In the other 6 cases there were various degrees of associated aortic insufficiency ranging from minimal and clinically insignificant to moderate, the prime lesion being the stenosis. In this latter group there was one death due to cardiac arrest with ventricular fibrillation occurring thirty minutes after completion of surgery. Although an admittedly poor risk case, the patient had withstood the procedure quite well. Table I.

TABLE I. UNIVALVULAR AORTIC LESIONS

LESION	NO. OF CASES	DEATHS			INSUFFICIENCY		FOLLOW-UP					ANGINA	
		EARLY	LATE	OPERATIVE	PREOP.	P.O.	WORSE	UNCHANGED	SLIGHTLY IMPROVED	IMPROVED	MARKEDLY IMPROVED	PREOP.	P.O.
Pure aortic stenosis (AS)*	7	-	-	-	-	2[B]	1[C]	-	1[D]	4	1	3	1
Essential aortic stenosis with asso. insuf. (AS, ai)**	6	1[A]	-	-	4	1	1[E]	-	-	3	1	1	-
Total	13	1			4	3	2		1	7	2	4	1

*Two cases: mitral valve explored; one at previous operation one and one-half years ago.

**Five cases: mitral valve explored.

[A]Cardiac arrest and ventricular fibrillation occurred thirty minutes after surgery was completed. Woman, aged 51, severe risk, chronic congestive failure unrelieved by intensive therapy. Mitral valve explored, normal. No tolerance to exercise. Myocardial failure one and one-half years. Autopsy: two aortic commissures, well opened. Note Fig. 2, Case E. S.

[B]P. P., a girl, aged 14, and W. E. I., a boy, aged 16, with congenital valvular aortic stenosis; both definitely improved, but with moderate and slight degrees of AI produced, respectively. Jetlike aortic systolic thrill changed to diffuse thrust; noted at surgery. (Eight and one and one-half months postoperatively, respectively.)

[C]R. N., a man, aged 30, three months postoperatively, had been in myocardial failure one year; now worse with sinus rhythm changed to auricular fibrillation; now requires diuretic therapy; has febrile illness believed by his physician to be either active rheumatic fever or subacute bacterial endocarditis. Blood pressure 110/60.

[D]J. D., a man, aged 67, highly calcareous valve with rapidly progressive myocardial failure preoperatively for six months. Slightly improved. Three and one-half months postoperatively.

[E]L. M., a man, aged 44, four months postoperatively, has more congestive failure with apparent recurrent rheumatic flare-up. There is no angina, heart size is diminished, no vertigo, normal sinus rhythm remains. Blood pressure 120/70.

TABLE II. MULTIVALVULAR LESIONS (AORTIC AND MITRAL)

LESION	SURGERY: AORTIC COMMISSUROTOMY	SURGERY: MITRAL VALVE: COMMISSUROTOMY	SURGERY: MITRAL VALVE: COMMISSURORRHAPHY	SURGERY: MITRAL VALVE: BOTH	DEATHS: EARLY	DEATHS: LATE	DEATHS: OPERATIVE	INSUFFICIENCY: PREOP.	INSUFFICIENCY: P.O.	FOLLOW-UP: WORSE	FOLLOW-UP: UNCHANGED	FOLLOW-UP: SLIGHTLY IMPROVED	FOLLOW-UP: IMPROVED	FOLLOW-UP: MARKEDLY IMPROVED	ANGINA: PREOP.	ANGINA: P.O.
AS-MS																
AS, MS	13	12[A]	–	–	2[B, C]	1[D]	–	–	–	–		–	6	4	–	–
AS, ai, MS	3	3	–	–	–	–	–	3	1[E]		1[F]	–	1	1	–	–
AS, ai, MS, mi	2	1[G]	–	–	–	–	–	2	–	–	–	–	1	1	–	–
AS, MS, mi	1	1	–	–	–	–	–	–	–	–	1[H]	–	–	–	–	?
Total	19	17			2	1		5	1		2		8	6		1?
AS-MS-MI																
AS, MS, MI	2	1[I]	–	1	–	–	1[I]	–	–	–	–	–	1	–	–	–
AS, ai, MS, MI	3	2	–	1[J]	1[J]	–	–	3	1	–	–	1	1	–	1	–
Total	5	3		2	1		1	3	1			1	2		1	
AS-MI																
AS, MI	2[L]	–	1	–	–	–	–	–	–	–	–	–	2	–	2	–
AS, ai, MI, ms	1	–	1	–	1[K]	–	–	1	–	–	–	–	–	–	–	–
Total	3		2		1			1	–		–	–	2		2	
AI-MS																
as, AI, MS	1	1	–	–	–	–	–	1	–	–	–	–	1	–	–	–
AI																
as, AI, mi	1	(also transaortic Nylon tamponage of the aortic valve; exploration of the mitral valve)														
Total	29	21	2	2	4	1	1	10	4		2	1	14	6	3	–

6 cases: died
20.7% mortality

21 cases:
72.4% improved

AS = Aortic stenosis.
AI = Aortic insufficiency.
MS = Mitral stenosis.
MI = Mitral insufficiency.
as, ai, ms, mi—minimal to slight degrees of same.

[A]In the case of I. M. B., a woman, 42 years of age, although a moderate degree of mitral stenosis was present, exploration was not attempted due to an extreme degree of dense pleuritic and pericardial fibrosis. There was an old heavily scarred and fixed left upper lobe, representing old and questionably still potentially active tuberculosis. Treatment eight years previously had been artificial pneumothorax.

This was the first case (April 3, 1952) in which the new instrument was used. The aortic valve was well dilated, changing the jetlike aortic thrill to a more diffuse thrust. Now eight months postoperatively, there is improvement, no coronary insufficiency, nor signs of myocardial failure.

[B]D. P. R., a man, 49 years old, died two and one-half hours postoperatively because of bleeding from the surgical incision in the left ventricle. This wound of entrance for the aortic dilator had been purse-stringed in the usual manner with no untoward accident. Interrupted closing atraumatic sutures were used in the heart with apparent hemostasis. There had been excellent recovery, but the true nature of the subsequent downhill course was not suspected until some fifteen minutes before death. Great efforts were put forth in an attempted revival. The aortic valve proved to be highly calcareous and had two commissures well opened, the anterior and the left posterior. Note Fig. 2. The operation occurred Dec. 23, 1952, and was the last of this series.

[C]H. W., a 41-year-old man, died on the seventh postoperative day due to massive pulmonary embolism. A soft antemortem thrombus was found in the right auricular appendage. It was felt that this was the site of origin of the pulmonary embolus. The anterior and left posterior aortic commissures were well opened. Immediate postoperative course had been one of definite improvement. (Fig. 2.)

[D]Mitral insufficiency produced in this combined case with recurrent pulmonary edema thereafter and death four and one-half months postoperatively. (Case of J. M., a 37-year-old man.) The anterior and posterior aortic commissures were well opened. (Fig. 2.)

[E]The aortic insufficiency remaining was considered insignificant (E. R. W., a woman aged 33 years).

[F]B. K., a woman, 44 years old, had a very significant degree of mitral stenosis and a moderate degree of aortic stenosis. There was a minimal to insignificant aortic insufficiency. There was a rheumatic history with recurrent myocardial failure for several years, and a two to three plus enlargement of the heart. The pulse pressure averaged 60 postoperatively, having been 47 preoperatively. However, at present the blood pressure is 122/76.

At surgery, following an instrumental mitral commissurotomy, the blood pressure fell to zero, the aortic systolic thrill having at first increased in intensity. The chest was rapidly closed and the patient was turned to the supine position. A re-entry was made anteriorly some thirty minutes later, and the aortic commissurotomy was performed with no difficulty. Postoperatively, there was a transient paralysis of the diaphragm, and a moderately severe degree of hysteria and neurosis lasting several days.

[G]M. Gn., a 46-year-old woman, had a calcified aortic valve with stenosis. The mitral valve was leathery, rolled, slightly stenosed with an estimated 1¼ finger sized opening and some 5 c.c. of regurgitation. It was decided to do nothing to this valve. There was a definite "crack" audible when the aortic valve was opened. At present, eight and one-half months later, the exercise tolerance is a good ten times better. This was primarily an aortic stenosis case, and the patient is listed as markedly improved.

[H]F. D., a woman, aged 46 years, with a combined stenosis of the aortic and mitral valves, also had an estimated 8 c.c. of regurgitation through the mitral valve. A 1 finger sized mitral stenosis was opened to 2 fingers by digital commissurotomy, resulting in an estimated 10 c.c. of regurgitation. Then, the aortic valve was widely opened, changing the aortic thrill from a jetlike to a diffuse type.

Preoperatively, this patient had had six months of severe myocardial failure, the last three months before surgery giving only a fair response to intensive medical therapy. There had been present a significant degree of essential hypertension for several years.

At surgery, on the induction of anesthesia, there was a cardiac arrest. Immediate cardiac massage was instituted with recovery. The double valve operation then was rapidly performed with no difficulty. This patient is listed as unimproved at the end of two months. The blood pressure is 210/110. There is a slight coronary insufficiency and some mild congestive failure. The aortic and mitral murmurs are reduced in intensity. There is a slight decrease in heart size.

[I]G. D. W., a man, aged 51 years, had a moderate degree of mitral stenosis with an estimated 20 c.c. of regurgitation through a leathery mitral valve. A small anterolateral mitral commissurotomy immediately caused more mitral insufficiency. It was believed best to open the aortic valve as soon as feasible. This was accomplished, but the purse-string suture in the exceptionally soft and friable ventricular myocardium tore through the heart wall with a severe hemorrhage following. There was an immediate fall in the blood pressure and cardiac arrest. Ventricular fibrillation ensued, and all attempts at recovery failed. Autopsy revealed a definite myocardial degeneration. There had been a history of angina for five years. Two of the aortic commissures were well opened. Specimen was not available for photography.

[J]R. F., a woman, aged 25 years, had advanced mitral stenosis, a moderate degree of mitral insufficiency, a moderate amount of aortic stenosis, and slight aortic insufficiency. Mitral commissurotomy and commissurorrhaphy were performed, and an aortic commissurotomy proceeded well. There was found at surgery a severe degree of pleuritic and pericardial adhesive change, with an almost acute inflammatory appearance. Unfortunately, no biopsy of this was obtained. She died on the twenty-eighth postoperative day after a steady downhill course terminating in congestive failure, jaundice, prolonged febrile course, and auricular fibrillation. Several transfusions, antibiotics, ACTH were used along with extended medical efforts.

as, al, ms, ml—minimal to slight degrees of same.

There had been a moderate degree of aortic insufficiency produced in a noncalcified valve. It was postulated that a rheumatic pericarditis, along with the physiologic load of the added aortic insufficiency and possibly surgically produced mitral stenosis resulted in progressive myocardial failure. Autopsy was not permitted. Biopsy of the auricular appendage demonstrated active acute rheumatic endocarditis.

[8]L. K., a 42-year-old woman, had essentially mitral insufficiency with a moderate degree of aortic stenosis, and some associated aortic insufficiency and mitral stenosis. Mitral stenosis was produced by too tight a commissurorrhaphy. Death occurred on the second postoperative day from pulmonary edema.

[1]K. B., a man, 40 years old, who, because of the evidence of severe myocardial damage, was operated on before the procedure of mitral commissurorrhaphy had become established, had correction of only the sclerotic aortic valve. No attempt was made to correct the mitral insufficiency. Now, some ten months later, there is no longer a mitral systolic murmur, implying that relief of the aortic stenosis has resulted in a significant ventricular pressure readjustment.

RESULTS WITH MULTIVALVULAR LESIONS

There were 29 cases with multivalvular disease in which aortic commissurotomy was performed with an over-all mortality of 20.7 per cent. Of these, 13 had "pure" aortic and "pure" mitral stenosis; 6 had essentially aortic and mitral stenosis, but associated with minimal to slight degrees of aortic and/or mitral insufficiency. There were 5 cases of aortic stenosis, mitral stenosis, and mitral insufficiency, 3 of which had minimal to slight degrees of aortic insufficiency; and there were 3 cases of essentially aortic stenosis and mitral insufficiency, of which one had very slight degrees of both aortic insufficiency and mitral stenosis. There was one case of aortic insufficiency and mitral stenosis with a slight degree of aortic stenosis. And there was a case of aortic insufficiency with slight degrees of both aortic stenosis and mitral insufficiency.

Out of these 29 cases, there were 25 with significant mitral stenosis; 23 mitral commissurotomies were performed. Two of the 23 had simultaneous mitral commissurorrhaphy (commissural suturing) for significant associated insufficiency.[8]

Also, in the 3 cases of aortic stenosis associated with a significant degree of mitral insufficiency, two commissurorrhaphies of the mitral valve were performed.

One case of aortic insufficiency associated with slight degrees of aortic stenosis and mitral insufficiency, in which an aortic commissurotomy and a transaortic Nylon graft tamponage of the aortic valve were performed, along with mitral valve exploration, had definite improvement as a result. Table II presents the results in these 29 cases of multivalvular lesions. There was a 72.4 per cent improvement clinically by follow-up, and a 20.7 per cent mortality, including early and late deaths.

OPERATIVE AND POSTOPERATIVE COMPLICATIONS

Technical.—

a. Hemorrhage: Case G. D. W. Due to the purse-string suture in the ventricle cutting through the myocardium following dilatation of the aortic valve, hemorrhage occurred. Cardiac arrest and ventricular fibrillation with temporary return lasting some minutes occurred, but the patient eventually died on the operating table. Case of AS, MI, ms.

Case D. P. R. Hemorrhage occurred due to insecure closure of the ventricular wound. Apparently there was hemostatis at the end of surgery, but bleeding ensued upon patient's return to his room. He died two and one-half hours later. Unfortunately the situation was not suspected until too late. All efforts were of no avail. The patient had done well in the very immediate postoperative period.

b. Aortic Insufficiency: P. P. and W. E. I. were two patients with congenital aortic valvular stenosis. Moderate to slight degrees of aortic insufficiency were produced, respec-

tively. Both patients definitely improved. It is now eight and one-half months postoperatively, respectively. (It is believed that step-wise opening of the valve, about one-third of the way each time, and then checking for any resulting insufficiency might have limited this effect.)

R. F. had aortic stenosis, mitral stenosis, and a slight degree of aortic insufficiency. The latter was significantly increased by the aortic dilatation. Simultaneous mitral commissurotomy and commissurorrhaphy were performed. Death came twenty-eight days postoperatively from congestive failure, acute pericarditis, and jaundice. The aortic insufficiency, per se, was not of sufficient degree alone to cause death, but was probably contributory in the degree of congestive failure.

c. Mitral Insufficiency: Mitral insufficiency was produced in a combined case of aortic and mitral stenosis. Both valves were operated on. Recurrent pulmonary edema occurred thereafter with death four and one-half months later (Case J. M.).

d. Mitral Stenosis: In Case L. K. mitral stenosis was produced in a combined case of aortic stenosis, mitral insufficiency, and minimal mitral stenosis. Aortic commissurotomy and mitral commissurorrhaphy were performed, the latter probably too tightly. Death occurred on the second postoperative day in pulmonary edema.

e. False Passage of Guidewire Through the Interventricular Septum Out Into the Pulmonary Artery: Further attempts were unsuccessful; recovery was uneventful. Case C. R., not reported here. The aortic valve was highly calcareous. This was an earlier case which might have been successful had we used the guiding finger tip via the left auricular appendage. There was co-existent mitral stenosis, which was relieved by commissurotomy before the attempted aortic commissurotomy. The patient was a 43-year-old man. Only slight improvement occurred.

Conduction Irregularities.—

a. Ventricular Fibrillation: Case D. R. Ventricular fibrillation occurred just prior to closure of the chest after pericardium had been closed. Combined mitral and aortic commissurotomy had been performed. Suddenly there were many extrasystoles and a tremendous "jumping" and beating of the heart; then fibrillation. The pericardium was opened and massaged, and two 3 second shocks of 110 volt house line current with the defibrillator were administered. More massage was applied until there was full recovery. The intravenous procaine had been stopped two minutes prior to the onset of the fibrillation. The patient was a woman 33 years old.

Case E. S. I. The essential difficulty was aortic stenosis with a slight degree of aortic insufficiency. There was a noncalcareous valve. The mitral valve was explored and was shown to be normal; aortic commissurotomy was performed. Death occurred very suddenly, 30 minutes after surgery was completed. Immediate thoracotomy showed findings of arrest and ventricular fibrillation. Treatment included massage and mouth-to-mouth respiration. Within five minutes positive pressure oxygen and defibrillation was attempted. All efforts proved unsuccessful. Valves had been well opened as per autopsy findings. This patient presented the worst risk of any case here reported. There had been severe myocardial failure with congestive findings unresponsive to intensive medical therapy for nearly three months. The patient was a 51-year-old woman.

b. Cardiac arrest: Case F. B. The patient was a 30-year-old woman, who had cardiac arrest while mitral commissurorrhaphy was being completed. Cardiac massage and IV (intravenous) atropine brought the heart back. Mitral commissurotomy and aortic commissurotomy were then performed. There were very rapid runs of auricular fibrillation postoperatively, and a borderline schizophrenas was present for several days. Eventual recovery and control of rhythm achieved. The patient improved.

c. Auricular fibrillation: E. Sg., a 39-year-old woman, was in a shocklike state on the fifth postoperative day after an operation of combined mitral and aortic commissurotomy. Rapid IV digitalization and pressure oxygen aided in full recovery.

TABLE III. AORTIC COMMISSUROTOMIES
(APRIL 3, 1952 TO DEC. 23, 1952)

Essential Aortic Stenosis	13		
"pure" AS		J. D., R. H., F. M., P. P., W. E. I., W. Er., R. N.	7
AS, ai		E. Sl., A. W. B., L. M. (slight ai)	3
		W. I., W. C. (sl. to mod. ai)	2
		G. B. (mod. to signif. ai)	1
Essential aortic stenosis and mitral stenosis	19		
"pure" AS, "pure" MS		M. D., E. Sg., H. W., I. M. B., H. M., D. R., D. P. R., M. Gi., R. U., J. M., J. B., M. C., W. J.	13
AS, ai, MS		B. K., S. B., E. R. W.	3
AS, ai, MS, mi		M. Gn., H. S.	2
AS, MS, mi		F. D.	1
Essential aortic stenosis, mitral stenosis, and mitral insufficiency	5		
AS, MS, MI		G. D. W., F. B.	2
AS, ai, MS, MI		E. C., F. K., R. F.	3
Essential aortic stenosis and mitral insufficiency	3		
AS, MI		C. W., K. B.	2
AS, ai, MI, ms		L. K.	1
Essential aortic insufficiency and mitral stenosis	1		
as, AI, MS		J. A.	1
Essential aortic insufficiency	1		
as, AI, mi		R. C.	1
		42 cases 7 deaths 16.6% mortality	

Isolated aortics	13 with 1 death:	7.7% mortality 77.0% improved
Multivalvular cases	29 with 6 deaths:	20.7% mortality 72.4% improved

R. F. (see under aortic insufficiency) died twenty-eight days postoperatively in congestive failure, probably with recurrent rheumatic infection and pericarditis. Severe runs of auricular fibrillation postoperatively; normal sinus rhythm had been present preoperatively.

Other Complications.—

a. Retained Secretions: Five cases required bronchoscopy and one tracheotomy (particularly common in cases with pulmonary fibrosis, emphysema, or a very large left atrium pressing on the left main bronchus).

b. Pleural Effusion: There were seven cases, serous, which recovered.

c. Atelectasis: There was one case; bronchoscopy and forced coughing; the patient recovered.

d. Pulmonary Embolism: One patient, H. W., a 41-year-old man, died on the seventh postoperative day. He had had aortic and mitral commissurotomy. Ante-mortem thrombus, right auricular appendage had occurred.

e. Gastric Dilatation: There were two cases; recovery was uneventful.

COMMENTS ON TWO CASES NOT A PART OF THOSE HERE REPORTED

The mandatory relief of both valves in combined aortic and mitral stenosis is aptly shown in two such cases in which only the mitral valve was opened.

Death from ventricular fibrillation and cardiac arrest came in one case, J. I. R., a man aged 58 years, 24 hours postoperatively. At surgery, following relief of the mitral stenosis, the blood pressure fell to a level of 60 and the decision was against attempting the aortic commissurotomy. This was one of the earlier cases; today both valves would have

TABLE IV. FORTY-TWO OPERATIVE CASES—CLINICAL EVALUATION

	AORTIC STENOSIS (40 CASES)				AORTIC INSUFFICIENCY	
42 CASES:	ISOLATED OR SIGNIFICANTLY SO (AS)	PLUS MITRAL STENOSIS (AS, MS)	PLUS MS & MI (AS, MS, MI)	PLUS MI (AS, MI)	PLUS MS (AI, MS)	WITH INSIGNIFICANT AS & MI (AI)
	13	19	5	3	1	1
Sex: 23 male	11	5	3	2	1	1
19 female	2	14	2	1	-	-
Age: (average)	36.2 yr.	41.5	36.5	43.3	34	34
(range)	14-67 yr.	32-59	25-51	40-48		
20 cases: positive history of rheumatic fever	4	8	4	2	1	1
19 cases: strongly suspected origin of rheumatic fever	6	11	1	1		
Other origin 3 cases	2 (congenital)			1 (Rheumatic fever and subacute bacterial endocarditis)		
Duration of known murmur: (average; valve unspecified)	15 yr.	14½	20	17	27	24
14 cases in myocardial failure	4	6	1	3	-	-
Duration of clinical signs and symptoms	4.3 yr.	5.5	3	9	6 mo.	10 yr.
Duration of acute symptoms (average)	1.2 yr.	1.1	1	9 mo.	6 mo.	-
Calcification of aortic valve	9	7	2	2	1	-
Mortality (16.6%)	1	0	1	0		
Operative	0	2	1	1		
Early	0	1	0	0		0
Late	1	3	2	1	0	-
Per cent	7.7	15.8	40.0	33.3	-	-

been opened. The lack of blood pressure would have caused concern but no surgical refusal to proceed.

Another patient, R. I., a 49-year-old woman, had relief of a tight mitral stenosis, but because the aortic stenosis was considered only slight to moderate in degree, further procedure was surgically refused. Some five weeks later this patient returned in severe congestive failure, with the obvious findings of significant aortic stenosis. Attempts to get the case in condition for surgery were to no avail in spite of intensive medical care. Death came in congestive failure nearly seven weeks postoperatively.

Comment.—These two cases demonstrate in a very vivid way the physiologic significance of multivalvular heart lesions and their requirements for simultaneous repair once surgery has been decided upon as the course of therapy. They tend to justify the concepts of surgical treatment as outlined under "Concepts."

SUMMARY

1. The etiology and pathogenesis of aortic stenosis is reviewed, including autopsy findings in 6 cases of 42 aortic commissurotomies.

2. Previous efforts for the relief of aortic stenosis are discussed.

3. Confirmation of the soundness of the concept of aortic commissurotomy is presented.

4. The Bailey aortic "dilator" is described.

5. The present technique of aortic commissurotomy is outlined.

6. Forty-two cases of aortic stenosis, with and without other valvular disease, are presented; in each instance aortic commissurotomy was performed. There were included 21 cases with mitral commissurotomy simultaneously performed, 2 with mitral commissurorrhaphy, 2 with both mitral commissurotomy and commissurorrhaphy, and one with transaortic Nylon tamponage of the aortic valve.

7. Concepts and discussion of physiology and technique are given.

8. Preoperative evaluation and indications for aortic commissurotomy are considered.

9. There were seven deaths in 42 cases, a mortality of 16.6 per cent. The causes are divided as follows:

In 13 cases of isolated aortic stenosis, one death, 30 minutes postoperatively, was due to ventricular fibrillation; mortality of 7.7 per cent.

In 19 cases of aortic stenosis and mitral stenosis (17 mitral commissurotomies performed) there were three deaths; mortality of 15.8 per cent. These are accountable as follows: one two and one-half hours after surgery due to bleeding from the ventricular surgical wound; one on the seventh postoperative day from a massive pulmonary embolism due to thrombosis of the right auricular appendage; one four and one-half months postoperatively due to recurrent pulmonary edema following a significant mitral insufficiency produced at surgery.

In 5 cases of aortic stenosis, mitral stenosis, and mitral insufficiency (each lesion significant) there were 2 deaths. This small group, besides having aortic commissurotomy, included 3 mitral commissurotomies and 2 in which both mitral commissurotomy and commissurorrhaphy (mitral commissural suturing) were performed. Mortality is 40 per cent. One death occurred on the operating table, following relief of the valvular leions, due to a tearing through of the hemostatic ventricular purse-string suture, resulting in fatal hemorrhage. The other death, twenty-eight days postoperatively, was due to reactivation of rheumatic infection with pericarditis, congestive failure, and perhaps some degree of produced aortic insufficiency.

In 3 cases of aortic stenosis and mitral insufficiency (3 aortic commissurotomies and 2 mitral commissurorrhaphies) there was one death; mortality of 33.3 per cent. This fatality occurred on the second postoperative day due to pulmonary edema probably resulting from a too tightly sutured mitral valve; a surgically produced mitral stenosis.

10. Of the isolated aortic valve cases, 77 per cent are improved. Of the multivalvular cases, 72.4 per cent are improved.

11. Operative and postoperative complications are discussed.

12. The theoretical concepts, the developmental experiences, the clinical and pathologic results of this work encourage us to hope that aortic commissurotomy for aortic stenosis will prove to be as permanently beneficial as mitral commissurotomy for mitral stenosis.

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CARDIOPULMONARY FUNCTION TESTS

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SINCE THE heart and the lungs function as a unit, it is often difficult to test them separately.

The main function of the heart and the lungs is to furnish a continuous supply of oxygen and nourishment to all cells of the body and to remove the carbon dioxide and metabolic end products.

FUNCTION OF THE HEART

The heart functions as a pump with the atrioventricular septum acting as a piston. Function tests will therefore in the first place include determinations of the pump's output and pressures in the heart and the big vessels.

The Cardiac Output.—The cardiac output is determined according to Fick's formula, $\frac{\text{Oxygen consumption ml./min.}}{\text{The arteriovenous } O_2 \text{ diff. ml./L.}}$. The error of the method, however, may be as much as 15 per cent in determinations at rest. To this analytic error a physiologic variation of the minute volume at two different investigations in the same subject must be added. The determination of cardiac output is, therefore, of limited value in evaluating the result of a valvulotomy. Only if the increase is more than 15 per cent may it be ascribed to the operation.

Furthermore it is difficult to compare the cardiac output at rest and during effort. If the patient at rest is anxious for a strange investigation a considerable increase of the cardiac output may result. Later on, when the patient is concentrating on limited work the cardiac output may have decreased. This decrease then cannot be ascribed to the exercise. Investigation during basal conditions is most important. Still, it is difficult to compare cardiac output at different occasions in the same subject.

The Arteriovenous Oxygen Difference.—The arteriovenous oxygen difference is a more reliable test of an adequate circulation. On two consecutive determinations in seven subjects with an interval of twenty to thirty minutes a standard deviation of 5.6 per cent was found in differences averaging 35 ml. per liter (Werkö, Berseus, and Lagerlöf, 1949). The technical errors will play a lesser role in cases with a higher arteriovenous oxygen difference. Values exceeding 50 ml. per liter at rest and 69 ml. per liter on effort are regarded as pathologic (Hickam and Cargill, 1948; Riley and his associates, 1948; and Dexter and his associates, 1951).

In cases of mitral stenosis grouped according to the New York Heart Association the following values were found (Eliasch, 1952):

From Sabbatsberg Hospital, Surgeon-in-chief, Professor C. Crafoord.
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	NO.	ARTERIOVENOUS O_2 DIFFERENCE
I. No symptoms II. No symptoms at rest Moderate symptoms on effort	23	38.0 ± 1.42
III. No symptoms at rest Pronounced symptoms on effort	22	52.6 ± 1.79
IV. Symptoms at rest	9	59.9 ± 3.47

Calculation of Shunts.—Calculation of the blood flow through the aorta and pulmonary artery and through the different shunts according to Dexter and Bing in cases of congenital malformations of the heart and big vessels will only give approximations of a very limited clinical value.

Pressure Measurements.—Pressure measurements are of the highest clinical importance. In cases with septal defects and pulmonary stenosis pressure measurements are of determining diagnostic value. Cases of mitral stenosis may be grouped according to the pressure in the pulmonary artery at rest and during effort. Such a classification is more objective and probably of more value than the one by the New York Heart Association.

A mean pressure of more than 6 mm. Hg in the right auricle is considered a sign of right heart failure.

Mean pressures exceeding 20 mm. Hg in the pulmonary artery are regarded as pathologic.

Regarding the pulmonary capillary pressure many unsolved problems remain. Dog experiments performed by Dexter suggest that the capillary pressure is almost identical to the pressure in the pulmonary veins. In cases with a high pressure in the pulmonary artery, however, this pressure may influence the capillary pressure to a certain degree. Therefore, it is not possible to decide with certainty from the form of the capillary pressure curve if a systolic pressure rise in a case of mitral stenosis with sinus rhythm is due to mitral regurgitation or a transmission of the pulmonary arterial pressure wave or to a summation of both factors.

The pulmonary capillary pressure curve does not help us to evaluate the degree of insufficiency complicating the mitral stenosis.

The pulmonary vascular resistance is calculated as follows: $\frac{\text{Pulmonary arterial mean} - \text{capillary mean pressure}}{\text{Cardiac output ml. per second}} \times 1{,}332$. Values exceeding 200 units are considered pathologic. The degree of pulmonary arteriolar resistance will express the change in the dynamics of the lesser circulation due to the mitral stenosis and causing the clinical symptoms.

During the progressive course of a mitral stenosis an increased pulmonary arteriolar resistance is developed. This increased resistance will prevent the right ventricle from pumping more blood into the lungs than the left heart can handle, thus preventing pulmonary edema.

How much pressure measurements and arteriolar resistance determinations will influence the indications for surgery can only be shown by future experience.

The pulmonary capillary pressure curve is of no conclusive help in the determination of the degree of mitral insufficiency complicating the stenosis.

The determination of the degree of complicating insufficiency before as well as after the valvulotomy is of utmost importance. Several laboratories are working on a technique for direct measurements in the left auricle along the following lines: 1. Retrograde catheterization from the aorta is technically impossible in most instances. 2. Introducing a needle directly into the left auricle through the anterior chest wall via the ventricle is feasible in dogs. 3. Direct puncture of the left auricle through the left main bronchus during bronchoscopy has been reported in ten cases. 4. I have used a method in which I introduce a needle paravertebrally directly into the left auricle from the right side.* Simultaneous pressure measurement in the pulmonary capillaries and left auricle shows that the pressure curve in the pulmonary capillaries closely reflects the pulse pattern in the left auricle in many cases (Fig. 1). The left auricular pressure curve seems, however, to have a more typical pattern in stenosis and insufficiency. We therefore hope that the left auricular pressure curve may be of help in evaluating the degree of insufficiency complicating a case of mitral stenosis.

The pulmonary capillary pressure is normal in cases of chronic pulmonary disease in spite of a high pressure in the pulmonary artery. A normal capillary pressure and a high pulmonary arterial pressure are also found in cases of Eisenmenger complex, that is tetralogy of Fallot without the pulmonary stenosis.

Having the patient breathe a gas mixture of about 10 per cent oxygen may produce symptoms and ECG changes in cases with coronary insufficiency.

The exercise test was introduced to measure the relationship between the pulmonary ventilation and gas exchange at rest and during exercise. Normal individuals show an increase in the oxygen uptake per liter of ventilation during effort. In patients with a mechanical obstruction to blood flow in the pulmonary artery, that is a pulmonic stenosis, the obstruction prevents a proportionate increase in pulmonary blood flow. The oxygen uptake per liter ventilation will therefore decrease during exercise (Bing). In Bing's experience 80 per cent of all patients with pulmonic stenosis showed a fall in the oxygen consumed per liter of ventilation. The value of oxygen consumed also fell in most patients with complete transposition of the great vessels.

PULMONARY FUNCTION

The main function of the lungs is a continuous exchange of oxygen and carbon dioxide between the alveoli and the pulmonary capillaries. The result of this gas exchange will immediately be visible in the oxygen and carbon dioxide tension of the arterial blood (provided cardiovascular anomalies can be excluded). Therefore an investigation of the arterial oxygen and carbon dioxide tension is important in the determination of the pulmonary function

*A detailed description of the technique will appear in *Annals of Surgery* (Björk, Malmström, and Uggla).

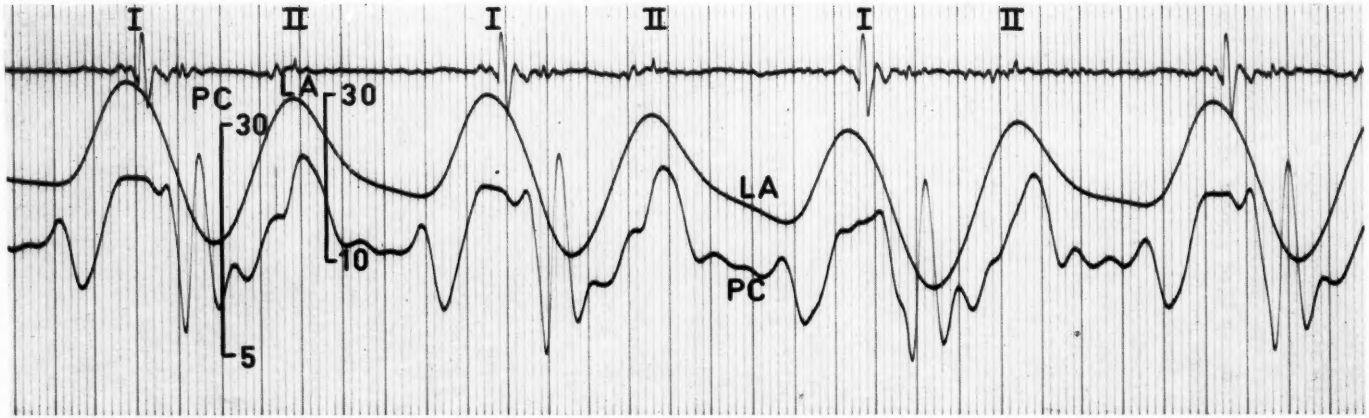

Fig. 1.—Simultaneous pressure measurement in left auricle (LA) and pulmonary capillaries (PC) in a case of mitral stenosis with some degree of regurgitation. The pulmonary capillary curve reflects the pulse pattern in the left auricle. The left auricular curve, however, is better configurated. I and II = the first and second heart sounds.

of patients. The following three functions are important factors in the gas exchange in the lungs. See Fig. 2.

(1) Ventilation, (2) Diffusion, and (3) Circulation—these three functions will be discussed separately. The result of the gas exchange, however, is visible in the arterial oxygen and carbon dioxide tension.

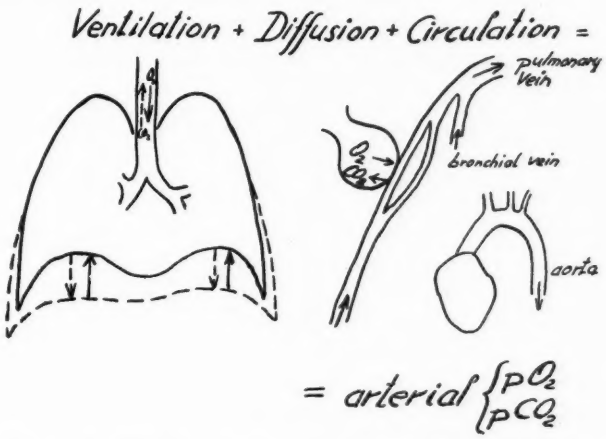

Fig. 2.—Diagram of the pulmonary function. The result of the gas exchange is visible in the arterial oxygen and carbon dioxide tensions.

To evaluate smaller variations in the oxygen contents of arterial blood the oxygen tension has to be determined. In 1951, Comroe wrote that blood leaves hyperventilated alveoli with practically no increase above normal arterial oxygen content. This opinion is based on investigations of the oxygen saturation according to Van Slyke and Neill. Therefore, it was most interesting to find a pronounced increase of the arterial pO_2 during hyperventilation. In healthy individuals there was an increase of pO_2 from 86 to 109 mm. Hg during hyperventilation while the oxygen saturation did not show any significant increase, see Table I. As such a pronounced increase in arterial oxygen tension is found during voluntary hyperventilation in healthy individuals, patients with pulmonary disease are now investigated in the same manner. It has been found so far that patients with no pulmonary reserve cannot increase their arterial oxygen tension during thirty seconds of hyperventilation. I therefore believe that the increase of arterial oxygen tension during voluntary hyperventilation is a good expression for the pulmonary reserve.

TABLE I. THE INFLUENCE OF HYPERVENTILATION ON ARTERIAL OXYGEN TENSION (pO_2) AND SATURATION ($\%O_2$) IN HEALTHY INDIVIDUALS

	NO. OF CASES	pO_2 MM. HG	NO. OF CASES	% O_2
Rest	29	86 ± 1.1	27	94.0 ± 0.4
Hyperventilation	21	109 ± 1.6	15	94.3 ± 1.0
Difference		23 ± 2.0		0.3 ± 1.1
		$P < 0.001$		$P > 0.05$

The explanation of the big difference in the oxygen saturation and the oxygen tension determinations is to be found in the dissociation curve of the oxyhemoglobin which is very flat at high tensions. See Fig. 3.

Variations in the amount of physically dissolved oxygen will hardly be detected by the saturation method but a significant difference will be found in the oxygen tension.

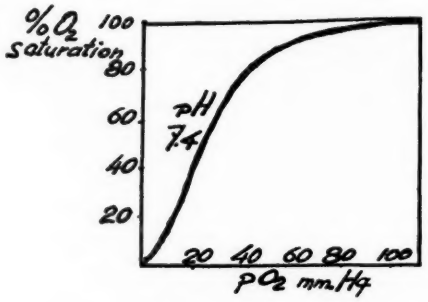

Fig. 3.—Diagram of the oxyhemoglobin dissociation curve, which is very flat at high tensions.

The sensitivity of the oxygen tension method is clearly demonstrated during heart catheterization. Blood was withdrawn from the pulmonary capillaries in sixteen patients and the mean pO_2 was 104.8 mm. Hg at the same time when blood from the brachial artery showed a mean pO_2 of only 77.6 mm. Hg. At the oxygen saturation determination no significant difference was found, see Table II.

TABLE II. A COMPARISON BETWEEN ARTERIAL OXYGEN TENSION (pO_2) AND SATURATION ($\%O_2$) IN BRACHIAL ARTERY AND PULMONARY CAPILLARIES IN 6 PATIENTS

	pO_2 MM. HG	% O_2
Brachial artery	86.4	93.4
Pulmonary capillaries	105.8	94.2

This high pO_2 value may be used to test whether or not the catheter is in the correct position for reading the pulmonary capillary pressure (Malmström and Michas).

The following factors contribute to the very high pO_2 found in the pulmonary capillaries: 1. The slow withdrawal of blood from the pulmonary capillaries will provide a longer equilibration time between alveoli and the capillaries. 2. As the circulation to this area is obstructed by the catheter, pO_2 in these alveoli will be higher than in the surrounding lung parenchyma. 3. The bronchial veins draining into the pulmonary veins will decrease pO_2 in the brachial artery compared to pO_2 in the pulmonary capillaries.

It is difficult to estimate the physiologic value of oxygen tension determinations in the pulmonary capillary blood as the two first mentioned factors are artefacts.

From this comparison of oxygen saturation and oxygen tension during hyperventilation and in pulmonary capillary blood, it is evident that only the oxygen tension of arterial blood is of value as a test of pulmonary function.

We use the technique of Riley, Proemmel, and Franke. The error of the method has been estimated by double determinations to 1.7 mm. Hg or 2.5 per cent of the average when single determinations are made, and to 1.2 mm. Hg or 1.8 per cent of the average when double determinations are performed.

Normal Values.—Twenty-nine healthy individuals have been examined. The mean value of arterial pO_2 was: 86.3 ± 1.08 mm. Hg (S.D. = 5.8; range 71.5 – 100.1 mm. Hg). Values below 70 mm. Hg are considered pathologic. (Mean value – 3 × S.D. = 68.9 mm. Hg.) Then an oxygenating insufficiency is diagnosed provided a right to left shunt can be excluded.

A patient with an oxygenating insufficiency has no cardiorespiratory reserve. Still, he may perform a limited amount of work.

Smaller and localized changes in the lungs will not decrease the arterial pO_2. Even patients with more pronounced pulmonary disease as well as patients who have had a pneumonectomy may have a normal arterial pO_2, provided the remaining lung tissue has a normal function.

It has been suggested (Comroe) that in borderline cases with a small cardiorespiratory reserve an oxygenating insufficiency should be more pronounced during effort. This has, however, not been the case in 29 normal persons and 108 patients with pulmonary disease where the arterial pO_2 has been investigated at rest and during effort.

1. In normal cases no difference is found in the arterial pO_2 at rest and during effort, see Table III.

TABLE III. OXYGEN TENSION (pO_2) AT REST AND DURING EFFORT IN HEALTHY INDIVIDUALS

	NO.	MEAN	S.D.
pO_2 at rest	29	86.3 ± 1.08	5.8
pO_2 during effort	19	85.4 ± 1.68	7.3

2. No decrease is found in the arterial pO_2 during effort in a group of thirty-four cases with oxygenating insufficiency at rest (pO_2 rest $<$ 65 mm. Hg). pO_2 rest $<$ 65 mm. Hg; n = 34; $\frac{\text{Difference } pO_2 \text{ rest} - pO_2 \text{ effort}}{-0.2 \pm 2.2,\ P > 0.05}$.

3. No decrease in arterial pO_2 during effort is found; either the maximal breathing capacity (MBC) is high or low, see Table IV.

TABLE IV. DIFFERENCE BETWEEN THE ARTERIAL OXYGEN TENSION (pO_2) DURING EFFORT AND AT REST

MEN	NO.	DIFFERENCE pO_2 EFFORT – pO_2 REST
MBC $>$ 95	25	–0.3 ± 1.9; P $>$ 0.05
MBC $<$ 56	19	–7.6 ± 4.4; P $>$ 0.05

4. If the ventilatory reserve $\left(100 - \frac{\text{minute ventilation during effort} \times 100}{\text{maximal breathing capacity}}\right)$ is high no correlation is found with the arterial pO_2 during effort. In cases with a very low ventilatory reserve ($<$ 50 per cent), however, a tendency to decreased arterial pO_2 during effort is noticed ($0.05 > P > 0.01$), see Table V.

TABLE V. COMPARISON BETWEEN INFLUENCE OF EFFORT ON ARTERIAL OXYGEN TENSION IN CASES WITH HIGH AND WITH LOW VENTILATORY RESERVE (100 – MV EFFORT/MBC)

	NO.	DIFFERENCE pO_2 EFFORT – pO_2 REST
100 – MV effort/MBC > 70%	47	0.002 ± 1.2; P > 0.05
100 – MV effort/MBC < 50%	23	–7.4 ± 2.8; 0.05 > P > 0.01

5. No decrease of the arterial pO_2 during effort was found; either the residual capacity was < 30 per cent or > 35 per cent of the total capacity.

It can therefore be concluded that during effort no significant change of the arterial pO_2 is found; either the pulmonary reserve is good or bad. The explanation to this finding is that the patients by a hyperventilation during effort can keep the arterial pO_2 at a fairly constant level. This hyperventilation is more pronounced in patients with a small pulmonary reserve.

The Carbon Dioxide Content of Arterial Blood.—The determination of the arterial pO_2 according to Riley and his co-workers is not superior to the manometric method of Van Slyke and Neill. Similar differences in the CO_2 content of arterial and pulmonary capillary blood were found at heart catheterization of six patients, see Table VI.

TABLE VI. COMPARISON BETWEEN CARBON DIOXIDE CONTENT IN BRACHIAL ARTERY AND IN PULMONARY CAPILLARIES IN 6 PATIENTS

	pCO_2 MM. HG	VOL. % CO_2
Brachial artery	35.6	45.4
Pulmonary capillaries	25.9	40.0

The very low CO_2 content in the pulmonary capillary blood must be ascribed an equilibration to ventilated, but not perfused, alveoli due to the occlusion of the heart catheter.

Also during hyperventilation similar differences of the CO_2 content are found by the two methods, see Table VII.

TABLE VII. INFLUENCE OF HYPERVENTILATION ON ARTERIAL CARBON DIOXIDE CONTENT STUDIED IN HEALTHY INDIVIDUALS

	NO.	pCO_2 MM. HG	NO.	VOL. % CO_2
Rest	29	35.0 ± 1.44	26	46.8 ± 0.73
Hyperventilation	21	16.4 ± 1.71	14	36.1 ± 0.73
Difference		18.6 ± 2.24		10.7 ± 1.03
		P < 0.001		P < 0.001

These findings clearly demonstrate the importance of taking the sample when the arterial needle has been in place for some time and the patient is breathing quietly.

The arterial CO_2 content in twenty-nine healthy individuals was found to be 35.0 ± 1.44 mm. Hg pCO_2, or 46.8 ± 0.73 volume per cent CO_2.

High arterial CO_2 content does not necessarily mean an insufficient CO_2 elimination. It may be due to primary chloride loss with compensatory retention of CO_2 as bicarbonate, that is alkalosis. If the arterial CO_2 content is elevated and the pH is decreased this indicates that the lungs can no longer eliminate CO_2 adequately and respiratory acidosis has occurred. As the blood

CO_2 content may be high or low in either alkalosis or acidosis, measurements of pH should always be performed to differentiate between these conditions.

Pulmonary insufficiency for CO_2 elimination is always accompanied by pulmonary insufficiency for oxygen uptake. On the other hand, the carbon dioxide content may be normal or low at times when an oxygenation insufficiency is present.

A normal arterial CO_2 content, therefore, does not exclude a very small pulmonary reserve. Then a hyperventilation of the alveoli will prevent the arterial CO_2 content to rise.

The arterial CO_2 content is higher in cases with an oxygenating insufficiency than in cases with arterial $pO_2 > 80$ mm. Hg; see Table VIII.

TABLE VIII. COMPARISON OF ARTERIAL CARBON DIOXIDE CONTENT IN CASES WITH HIGH AND LOW ARTERIAL OXYGEN TENSION

	NO.	pCO_2 MM. HG	NO.	VOL. % CO_2
$pO_2 > 80$	32	37.6 ± 1.5	22	44.9 ± 0.8
$pO_2 < 65$	34	44.5 ± 1.6	24	48.1 ± 1.2
Difference		6.9 ± 2.2		3.2 ± 1.4
		$0.01 > P > 0.001$		$0.05 > P > 0.01$

In cases with a high residual capacity and a low maximal breathing capacity no significant increased arterial CO_2 content was found although it was high in a few cases.

No significant change of the arterial CO_2 content during effort was found either in normal persons nor in patients with oxygenating insufficiency.

In cases with a decrease of the arterial $pO_2 > 10$ mm. Hg during effort an increased arterial pCO_2 was found. On the other hand, cases with an increase of arterial $pO_2 > 7.5$ mm. during effort showed a decreased pCO_2 at the same time, see Table IX. It is therefore evident that the influence of effort on the arterial CO_2 content is dependent on the degree of hyperventilation.

TABLE IX. COMPARISON BETWEEN INFLUENCE OF EFFORT ON ARTERIAL CARBON DIOXIDE CONTENT IN CASES WITH DECREASE AND INCREASE OF ARTERIAL pO_2 DURING EFFORT

DIFFERENCE EFFORT – REST	NO.	DIFFERENCE EFFORT – REST pCO_2 MM. HG	NO.	DIFFERENCE EFFORT – REST VOL. % CO_2
$pO_2 > -10$ mm.Hg	26	6.4 ± 2.4	24	–2.5 ± 1.5
$pO_2 > +7.5$ mm.Hg	22	–5.0 ± 2.4	14	–3.7 ± 1.2

Bronchospirometry.—Bronchospirometry will give information about the oxygen uptake and ventilation of each lung separately. According to Björkman 54 to 55 per cent of the oxygen uptake and ventilation occurs in the right lung in healthy individuals. The diminished oxygen uptake of the left lung is partly considered to be due to a longer left main bronchus. Baldwin and his associates consider the share of the right lung in oxygen uptake to vary from 50 to 65 per cent.

The diseased lung will show a relatively better function breathing 100 per cent oxygen, compared to the function when air is used.

If bronchospirometry is performed in the lateral position the oxygen uptake for the lower lung is increased. This is probably due to an increased blood flow through the lower lung and as well to an increased ventilation of the lower lung. This increased ventilation of the lower lung is due to the high position in the thorax of the lower diaphragm which has an extreme expiratory position facilitating a big inspiratory excursion.

This finding is constant and may be used as a function test (Steinman). If the diseased lung during bronchospirometry does not increase its oxygen uptake when the patient is in a lateral position with the diseased lung lowermost, this is considered to show a very small reserve function of the lung.

It is difficult to draw any conclusion from the absolute value of a lung's oxygen uptake per minute during bronchospirometry as the cardiac output may increase up to 50 per cent during the examination.

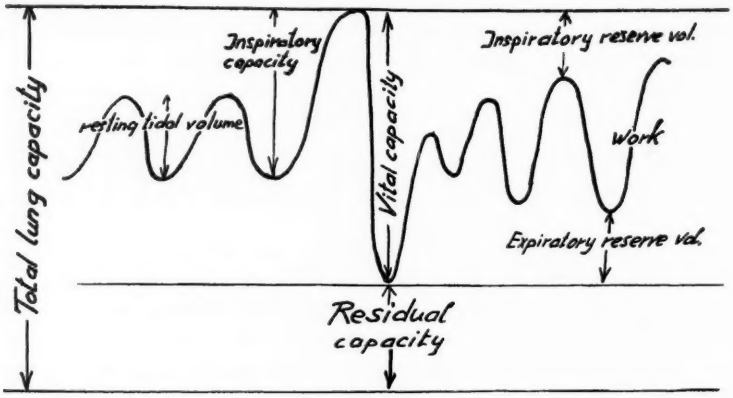

Fig. 4.—The terminology of the lung volume and its various subdivisions.

Bronchospirometry during effort has been studied by Björkman and Carlens. No further information is to be gained by this investigation as the share of the oxygen uptake by both lungs is the same at rest and during effort.

Changes in the pulmonary parenchyma and the bronchi as well as changes of the pleura decrease the function. In cases of unilateral parenchymal disease it is mainly the oxygen uptake which is diminished. In these cases the ventilation is never decreased to the same degree as the oxygen uptake. It may be possible to find cases with a pronounced decrease of the oxygen uptake of one lung at the same time that the ventilation is normal.

Bronchospirometry is the only method for studying the change in function of the operated lung and the compensatory change of the contralateral lung.

Through bronchospirometric investigations we have found a tremendous loss of function due to pleural disease. When an intrapleural pneumothorax is abandoned the re-expansion is often slight; it is mainly a shift of the mediastinum, an elevation of the diaphragm, and narrowing of the ribs that diminishes the pneumothorax space.

An extrapleural pneumothorax on the other hand will cause only a slight impairment of the function compared to an intrapleural pneumothorax. After a lobectomy about one-third of the total function will remain on the operated side. A segmental resection may not cause a significant decrease of pulmonary function. Decortication may improve the pulmonary function considerably.

Ventilation.—There has been a certain confusion regarding the terminology of the lung volume and its various subdivisions. A committee of respiratory physiologists has agreed upon the following terminology and definitions. See Fig. 4.

Vital capacity is the maximal volume of gas that can be expelled from the lungs by forceful effort following a maximal inspiration. This is the oldest lung function test. Volume is measured unrelated to time. It is an adequate measure of ventilation only if the outflow of air from the lungs is unimpeded. There is no correlation of vital capacity to emphysema or bronchial obstruction. The patient may have oxygenating insufficiency and a normal vital capacity. It is important to eliminate such an unsuitable test. A much more valuable test of ventilation is the maximum breathing capacity where volume per unit of time is measured.

Maximum breathing capacity is defined as the maximum volume of gas that can be breathed per minute. Usually the expiratory air during a 15 second period is measured. The best result is obtained at a respiration of between 40 to 70 per minute. Normal values for the maximal breathing capacity (MBC) vary somewhat in different laboratories with the resistance to breathing in the different spirometers used. In our laboratory healthy individuals from 14 to 62 years of age (mean 28.3 years) showed the following values (see Table X).

TABLE X. NORMAL VALUES FOR MAXIMAL BREATHING CAPACITY (MBC). AGED 14 TO 62

	NO.	MEAN	S.D.	RANGE
MBC men	18	127 ± 7.5	31.8	80-188 L./min.
MBC women	11	102 ± 4.9	16.2	82-130 L./min.

Healthy men between 50 to 70 years of age have a MBC of 60 to 140 liters per minute (mean 90) and healthy women in this older age group have an MBC of 50 to 100 liters per minute (mean 73.5) (Comroe).

MBC is usualy low in cases with emphysema due to the diminished elasticity and due to the extreme inspiratory position of the chest. If the MBC is compared to the arterial oxygen tension at rest a significant correlation is found in men (82 men, corr. coeff. 0.62 ± 0.07 $P < 0.001$) as well as in women (52 women, corr. coeff. 0.60 ± 0.086, $P < 0.001$). See Fig. 5.

If then the arterial oxygen tension in men with MBC $>$ 95 liters per minute is compared to the arterial oxygen tension in men with MBC $<$ 56 liters per minute a statistically significant decrease is found in the later group, see Table XI.

TABLE XI. COMPARISON OF ARTERIAL OXYGEN TENSION IN MEN WITH HIGH AND LOW MBC

MEN	NO.	pO_2 MM. HG
MBC > 95 L./min.	25	79.1 ± 1.6
MBC < 56 L./min.	19	65.1 ± 2.3
Difference		14.0 ± 2.8 P < 0.001

Thus a significant correlation between the MBC and the oxygenating function of the lungs has been found. These findings demonstrate the importance of MBC as a lung function test.

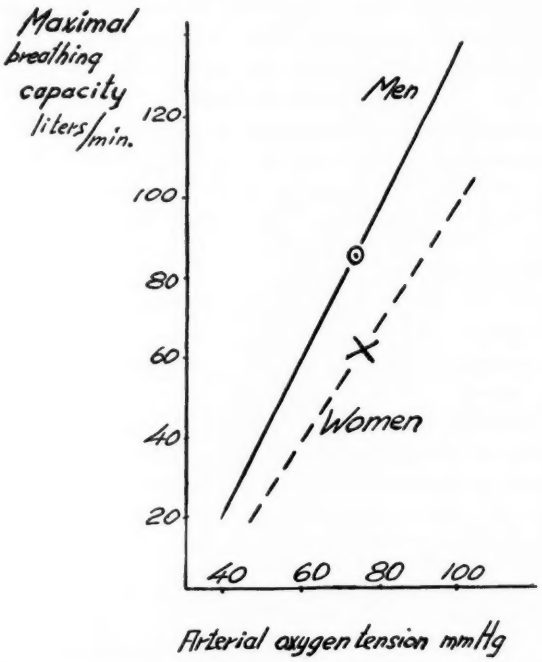

Fig. 5.—A significant correlation is found between the maximal breathing capacity and the arterial oxygen tension at rest.

Ventilatory reserve has earlier been defined as MBC minus minute ventilation at rest (Cournand and Richards). The breathing reserve index is:

$$\frac{\text{MBC} - \text{minute ventilation at rest}}{\text{MBC}} \times 100.$$

Ornstein and his associates have calculated the breathing reserve from the ratio $\frac{\text{MBC}}{\text{minute ventilation at rest}}$. Only Warring has earlier correlated the minute ventilation during walking to MBC as an expression for the breathing reserve.

It is, however, evident that the minute ventilation during rest is fairly constant; either the ventilation is good or bad. Therefore, these calculations of the breathing reserve in which the minute ventilation at rest is used are

without any value. In this ratio only the MBC will show any variation. The test will therefore give no further information apart from the MBC. In an investigation of seventy-three patients no correlation was found between the arterial oxygen tension and the ratio minute ventilation at rest/MBC, see Fig. 6.

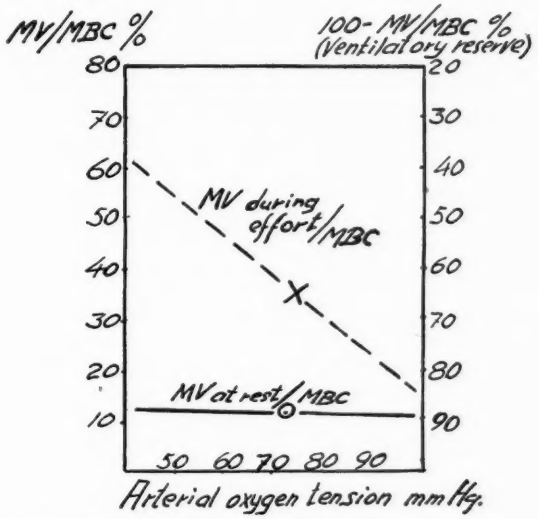

Fig. 6.—No correlation is found between the arterial oxygen tension and the ratio minute ventilation (MV) at rest/maximum breathing capacity. On the other hand a significant correlation is found between the arterial oxygen tension at rest and the ratio minute ventilation during effort/maximal breathing capacity.

In spite of a sincere desire to diminish the amount of pulmonary function tests we have introduced a new term, *ventilatory reserve,* calculated from the minute ventilation during effort (300 kilogram per minute on a bicycle ergometer). Ventilatory. reserve $= 100 - \frac{\text{Minute ventilation during effort} \times 100}{\text{MBC}}$

The ventilatory reserve was calculated from nineteen healthy individuals to $77 \pm 1.4 = \text{S.D.} = 6.2$; range = 86.5 to 65.5 per cent. Ventilatory reserve of less than 55 per cent is considered pathologic. The correlation between the ventilatory reserve and the MBC in men is good and the following values were obtained, see Table XII. The correlation between the arterial oxygen tension and the ventilatory reserve is also good. See Fig. 5, which shows the regression equation.

TABLE XII. COMPARISON OF VENTILATORY RESERVE (100 – MV EFFORT/MBC) AND ARTERIAL OXYGEN TENSION IN MEN WITH HIGH AND LOW MBC

MEN	NO.	100–MV EFFORT/MBC	NO.	pO_2 MM. HG
MBC > 95 L./min.	23	77 ± 3.0%	25	79.1 ± 1.6
MBC < 56 L./min.	17	41.4 ± 4.9%	19	65.1 ± 2.3
Difference		36.6 ± 5.7		14.0 ± 2.8
		P < 0.001		P < 0.001

Residual Air.—The residual capacity is the volume of gas remaining in the lungs at the end of a maximal expiration. Values of more than 35 per cent of the total lung volume are considered pathologic (Birath). In normal cases the values are reliable and duplicate residual air values often check within 3 per cent. In patients with emphysema and an uneven intrapulmonary mixing the method is less precise. An increased residual capacity is caused by hyperinflation of the lung and may be due to: (1) true emphysema, tearing of the alveolar septa, and reduction in pulmonary capillary bed; (2) obstruction to the airway, that is asthma; (3) compensatory overinflation of the lung following resection of lung tissue; (4) deformity of the thorax; (5) decreased elasticity of the lung in the older age group; (6) fibrosis and congestive heart failure.

In our cases with increased residual capacity we have found a decreased arterial oxygen tension (oxygenating insufficiency) and a decreased ventilatory reserve (ventilatory insufficiency).

An uneven distribution of the respiratory air with underventilation of the alveoli and hyperventilation of others may, depending upon the distribution of the pulmonary capillary blood flow lead to an oxygenating insufficiency. A comparably simple test of the intrapulmonary gas mixing has been devised by Cournand and his co-workers. The patient breathes oxygen for seven minutes and at the end of this period an alveolar gas sample is analyzed for nitrogen concentration which in normal cases may not exceed 2.5 per cent nitrogen. If the inspired oxygen is evenly distributed to all alveoli a very small amount of nitrogen remains after seven minutes. On the other hand, in badly ventilated alveoli the nitrogen concentration remains high and will show an abnormally high value of nitrogen in the final alveolar sample after seven minutes. This test is influenced by the lung volume, the effective tidal volume, and the frequency of breathing as well as by the distribution factor.

In cases with a small minute ventilation, too high a value may be obtained. On the other hand, a too low value may be obtained in cases with a pronounced emphysema due to hyperventilation. This shows the limitation of the method.

A more sensitive method is to use a rapid gas analyzer such as the nitrogen meter. In a group of patients 78 per cent were found to have an abnormal intrapulmonary mixing by this sensitive method compared to only 32 per cent when the Cournand test was used (Comroe and Fowler).

By bronchospirometry the determination of ventilation of each lung separately is determined. The determination of the maximal diaphragmatical movements will also give certain information of the ventilation of each lung.

Diffusion.—Oxygen is going from the alveolus with a high pO_2 through the alveolar membrane to the capillary where pO_2 is lower. The difference in pO_2 on each side of the alveolar membrane is less than 12 mm. Hg (Cournand and his associates). In these determinations blood from the brachial artery has been used. It is, however, necessary to realize that some of the bronchial veins drain into the pulmonary veins thus decreasing pO_2 in the arterial blood.

Furthermore some of the alveoli may have a poor ventilation. In view of these two facts it is highly probable that the difference in pO_2 in the alveoli and the pulmonary capillaries is very small indeed. Alveoli in different parts of the lung are ventilated differently. At rest there is probably a great variation of blood flow through different parts of the lung. It is probable that part of the pulmonary capillaries are not perfused when not needed. To test the diffusion the patient is breathing 100 per cent oxygen for ten to fifteen minutes. Provided no shunt exists the arterial blood should be fully saturated with oxygen when the diffusion is adequate. If the arterial blood then is unsaturated this may be due to diffusion impairment.

The Pulmonary Circulation.—In the pulmonary circulation the vascular resistance is relatively low at a high flow. The circulation time is, therefore, short. It will only take about two seconds for blood from the right ventricle to reach the pulmonary capillaries and about the same time to return to the left auricle. At rest the blood is passing the pulmonary capillaries in three-fourths of one second and during effort in only one-third of a second (Roughton, 1945).

The pulmonary arterial resistance is calculated in units:

$$\frac{\text{Pulmonary artery pressure} - \text{pulmonary capillary pressure}}{\text{Cardiac output ml./second}}$$

. Normally the pulmonary capillary pressure is less than one-half the capillary pressure in the systemic circulation. Therefore, a considerable safety margin against pulmonary edema exists.

In advanced cases of pulmonary disease emphysema and pulmonary fibrosis may cause a right heart failure, also called "cor pulmonale." It has been assumed that the destruction of the pulmonary capillaries in the lung should be the cause of the increased vascular resistance. If this were the case, the pulmonary capillary pressure should increase as well. In eighteen cases with pronounced pulmonary emphysema and fibrosis, however, Dexter found a normal pulmonary capillary pressure in spite of an increased pulmonary arterial pressure. The arteriolar resistance in the lung could be increased up to more than twenty times the normal value. In cases with an increased pulmonary arteriolar resistance a decreased cardiac output and an increased arteriovenous oxygen difference was found. The impairment of circulation in cases with advanced pulmonary disease is, therefore, considered to be related to the resistance to blood flow through the pulmonary arteriole (Dexter). On the other hand, Dexter did not find any correlation with the pulmonary arteriolar resistance and the maximal breathing capacity, the vital capacity, and the residual air.

In cases with chronic diseases causing a considerable reduction of the vascular bed and an increased pulmonary arterial pressure with a low cardiac output the prognosis is considered poor. In healthy individuals Cournand and his co-workers consider the cardiac output at rest may increase up to more than three times this value without any significant increase in pulmonary arterial pressure. Due to this big margin the pulmonary arterial pressure is found to be within normal limits in spite of a very advanced pulmonary disease.

During effort with an increased cardiac output a pathologic increase of the pulmonary arterial pressure will more easily be detected (Uggla).

An increased pulmonary arterial pressure is, however, no contraindication to a pulmonary resection. Such a high pressure may, however, in the future be of help in determination of the prognosis.

Carlens, Hanson, and Nordenström at the Sabbatsberg Hospital have developed these pressure measurements further by occlusion of one of the pulmonary arteries when the pressure in the other artery is determined at rest and during effort. The occlusion is achieved by filling a rubber balloon in the end of the double lumen catheter. Through the other opening of the catheter the pressure is measured centrally to the occlusion. This method is most valuable in cases where a pneumonectomy is planned and the diseased lung is considered to function at the same time as the function of the best lung is impaired.

With this method it is also possible to determine if a shunt mechanism exists, that is, if blood is perfusing poorly ventilated alveoli without being saturated by oxygen. Then an increased pO_2 in the arterial blood is found after occlusion of the pulmonary artery to this lung (Hanson).

It is also possible to get some information about the relative blood flow through the two lungs by bronchospirometry as the blood flow is proportional to the oxygen uptake.

If a lung or a lobe is completely atelectatic or destroyed we know that the shunt has been eliminated and practically no blood is passing the atelectatic or destroyed part of the lung. To determine the amount of blood passing an atelectatic or destroyed part of the lung I introduced the following method:

During operation blood samples are taken: 1. First a blood sample is taken from the aorta. 2. Then a blood sample is taken from one or both pulmonary veins when the vein is occluded centrally. 3. When both veins from the diseased lung are ligated a new blood sample is taken from the aorta. This blood sample may then be considered to represent blood from the pulmonary vein of the other lung. Then if the oxygen saturation in the pulmonary veins of both lungs as well as the oxygen saturation from the mixture of blood from both lungs (that is from the aorta) are known, it is easy to calculate the relative flow of blood through the two lungs.

In order to confirm these investigations I have during operation opened the pulmonary veins and measured the flow of blood per unit of time which has been collected when the pulmonary artery is intact but all bronchial vessels divided. The blood flow through an atelectatic or a destroyed lung has been found to be too small to be of any practical importance in the development of an oxygenating insufficiency.

CONCLUSIONS

The best of all pulmonary function tests is the operation itself.

We have in more than one hundred cases of impaired lung function performed a pulmonary resection combined with thoracoplasty in spite of a pronounced oxygenating insufficiency and ventilation insufficiency with a maxi-

mal breathing capacity of in some cases less than 20 liters per minute. We have not sufficient experience to give the minimum value of pulmonary function for the patients standing a pulmonary resection. Only one of our patients died in connection with the operation due to very poor pulmonary function. She had a pronounced ventilatory insufficiency after a bilateral pneumothorax. After the operation she regained consciousness but could not adequately oxygenate herself breathing spontaneously. Another patient died from oxygenating insufficiency three weeks after the operation. Three patients developed pulmonary edema in connection with the operation. A tracheotomy was immediately performed in order to diminish the dead space and improve the ventilation. These patients have no strength for coughing and the repeated aspirations of the bronchial tree through the tracheotomy contributed to the successful outcome in all three patients.

The immediate postoperative period after a pulmonary resection is therefore considered to be endured by practically all patients with a very poor pulmonary function.

We consider the following lung function tests ought to be done and in the order listed.

1. In cases of pulmonary tuberculosis an evaluation of all x-ray films from the very beginning of the disease will give the surgeon most important information about the pulmonary function.

2 The clinical investigation is important but will not necessarily give the desired information. A pronounced dyspnea at rest is a bad sign.

3. The ventilation is investigated by determination of the maximal breathing capacity (MBC) and ventilatory reserve $\frac{(100 - \text{MV effort} \times 100)}{\text{MBC}}$.

In most cases these investigations are sufficient for an experienced surgeon to determine if the pulmonary reserve of the patient is large enough for the proposed operation.

From a practical point of view it is not possible to do more investigations on cases with an adequate ventilation in most thoracic surgical units. Furthermore in most cases there exists a good correlation between the ventilation and the oxygen uptake.

4. In cases with ventilatory insufficiency the determination of the oxygen and carbon dioxide tension in the arterial blood ought to be performed. The determination is performed both at rest and during voluntary hyperventilation. The increase of arterial oxygen tension during hyperventilation may be regarded as an expression for the pulmonary reserve.

5. The residual capacity may give valuable information.

6. Bronchospirometry is most valuable test in many cases. Earlier it has been considered of utmost importance in cases where the best function has been found on the diseased side. This finding then often constituted a contraindication to operation on the best side.

According to our experience, however, the risk of postoperative atelectasis is very small if a partial resection is performed on the lung with the best

function. If an operation is performed on the best lung the patient is forced to ventilate the remaining lung parenchyma on the side of operation well enough to prevent an atelectasis. As an area of the lung with no or minimal function is resected the chances to stand an operation on the lung with the better function are very good.

Bronchospirometry should not be performed in tuberculous cases with big cavities and considerable expectoration. In cases with tuberculosis the bronchospirometry should, if possible, be performed in connection with the operation. In these cases we always use Carlens double lumen catheter for endobronchial anesthesia in order to prevent spillover. When the tube is introduced we perform the bronchospirometry immediately on the operating table and then continue with the resection.

7. The most physiologic way to determine if the patient will withstand a pneumonectomy is the occlusion of the pulmonary artery according to Carlens, Hanson, and Nordenström. This investigation is of value in cases where the remaining lung has an impaired function.

8. Pressure measurements in the pulmonary artery are of no help in determining the immediate operative risks. These investigations may, however, be of prognostic value.

We have yet a lot to learn about pulmonary function. It is our intention to use few but adequate function tests. We believe that with the aid of our present pulmonary function tests we may determine if a patient can withstand an operation or not, but we do not yet know the value of our function tests in regard to the future prognosis for the patient.

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LEIOMYOMA OF THE ESOPHAGUS

Report of a Case With Successful Enucleation of the Tumor

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TUMORS of the esophagus may be divided into those which are malignant and those which are benign. While the prognosis for malignant tumors is extremely bad, most of the literature has concerned itself with detailed descriptions of the anatomy, microscopic pathology, and surgical treatment of carcinoma of the esophagus. Benign tumors of the esophagus, on the other hand, are infrequent enough so that whenever a case is found either at the operating table or at the post-mortem examination it usually reaches the literature. In spite of this fact, when Garlick and Stegmaier summarized the literature up to January, 1951 there were less than twenty cases of benign tumors of the esophagus, and in most of these the lesion had been discovered at autopsy. In May, 1951, Myers and Bradshaw[3] reported that they were able to find only 39 surgically treated benign intramural tumors of the esophagus; to these they added three of their own. We have found two additional cases reported since that time.[2, 4] This paper adds still another case to those already reported; it is a case in which the tumor was diagnosed preoperatively and the lesion was successfully removed surgically.

CASE REPORT

S. R., a 37-year-old white woman, was first seen by one of us (S. O. K.) in 1948. At this time she complained of vague indigestion, right upper quadrant and epigastric pain, "bloating" shortly after meals, and some intolerance to fatty and fried foods. Most of these symptoms were of recent onset. The postprandial bloating had existed for at least ten years. The initial physical examination was normal except for a mild hypotension and the presence of a right lower quadrant appendectomy scar.

Electrocardiogram, blood count, and urinalysis were entirely normal. Fluoroscopic examination of the chest revealed no abnormal pulmonary or cardiac shadows. A barium swallow revealed some spasm at the distal end of the esophagus. No films of the esophagus were taken at this time. An oral cholecystogram demonstrated a poorly functioning gall bladder with many small radiopaque stones floating at a uniform level. In view of these findings, in December, 1948, a cholecystectomy was performed from which the patient made an uneventful recovery.

About one year following the cholecystectomy the patient began to complain once again of bloating shortly following meals. In addition there was also a sensation of giddiness, vertigo, and constipation. It was felt that many of these symptoms were due to a state of mild anxiety, and treatment was ordered in that direction. The symptoms remained about the same and once again a barium swallow and fluoroscopic examination revealed some spasm at the lower end of the esophagus. However, at this time there appeared to be some distortion of the normal pattern.

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The patient was admitted to the Albert Einstein Medical Center, Northern Division, for study. All studies other than those related to the esophagus were within normal limits. The roentgenologist interpreted the x-ray films as showing either a mediastinal mass encroaching upon the esophagus or an intramural esophageal tumor (Fig. 1). A slight hiatal hernia was also present. The patient refused esophagoscopy and was discharged, taking a mild antispasmodic and a fairly normal diet.

Fig. 1.—X-ray view of the lower esophagus taken in November, 1950, showing the deformity produced by the lesion.

Since the patient was very apprehensive about any operative procedure, it was decided to re-examine the esophagus roentgenologically at regular intervals. Two successive series of films failed to show any change in the contour or size of the mass (Fig. 2). Although the patient's symptoms were fairly well controlled by antispasmodics, episodes of nausea, bloating, and even vomiting occurred, particularly when she was under any emotional stress. In view of the fact that the latter group of symptoms appeared to increase in severity surgical intervention was again recommended and on Nov. 27, 1951, she was admitted to the hospital for operation.

Operation was performed on Nov. 29, 1951. With the patient under endotracheal anesthesia the left thoracic cavity was entered through a posterolateral incision, removing the eighth rib from the costal margin to the angle. Exploration of the mediastinum revealed a large tumor mass in the lower third of the esophagus, involving approximately two-thirds of the circumference of the organ. The mediastinal pleura over the lesion was incised and the esophagus mobilized. The outer wall of the esophagus over the lesion was incised and the tumor was completely enucleated without entering the lumen of the organ. While the outer muscular layer of the esophagus was somewhat attenuated producing a diverticular defect, it was felt inadvisable to resect any of this tissue. The wall was then reconstructed with several interrupted sutures of 00 chromic catgut, and the chest was closed in layers. A large catheter was placed in the pleural cavity for drainage.

The resected specimen consisted of a mass, irregular in shape, moderately soft in consistency, gray-white in color, beneath a thin transparent, fibrous, hemorrhagically discolored

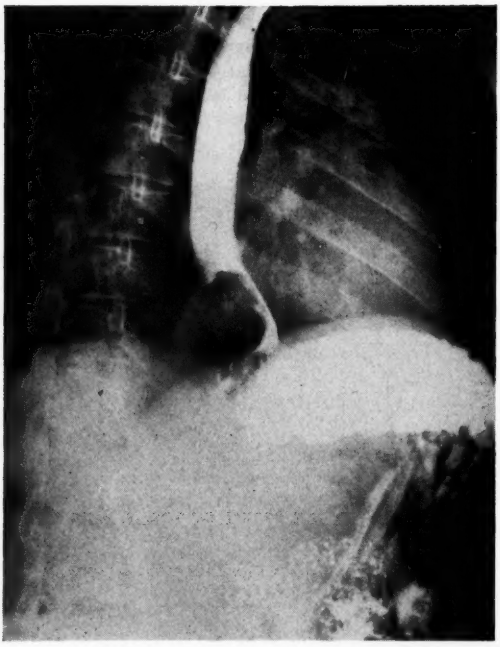

Fig. 2.—X-ray film taken one year later showing no apparent change in the appearance of the lower esophagus.

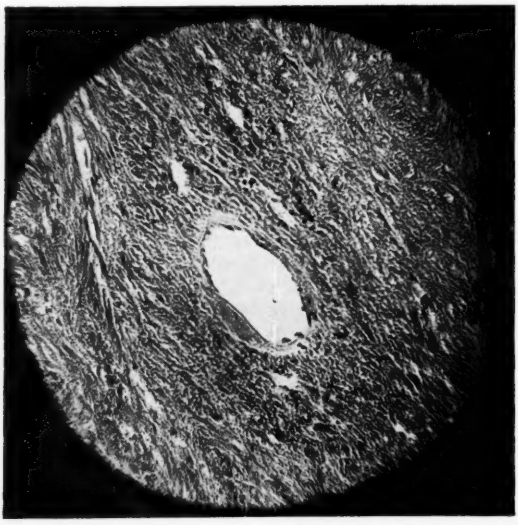

Fig. 3.—Microscopic section of the lesion. There is no evidence of malignancy.

capsule. It measured 8 by 4 by 4 cm. Its cut surfaces were homogeneously gray-white and whorled. Microscopic examination revealed the lesion to be a benign leiomyoma (Fig. 3).*

The postoperative course was entirely uneventful. Oral feedings were started on the third day and rapidly increased, so that the patient was discharged on the tenth day on a bland diet. The intercostal catheter drained for twenty-four hours and was removed on the third postoperative day. There was a moderate pleural effusion in the left pleural cavity, but this absorbed before the patient left the hospital. At no time was there any difficulty in swallowing and at no time during the postoperative course did the patient vomit. The day before the patient left the hospital an x-ray film of the lower esophagus was made, so that we might have a baseline study of the anatomy and function of the organ. This was con-

Fig. 4.—X-ray photograph taken immediately after operation showing the diverticular defect where the lesion had been removed.

sidered especially important in view of the marked redundancy of the wall following the enucleation of the tumor. The x-ray pictures made at this time showed no obstruction. There was a slight redundancy of the distal esophagus with the formation of a small dependent pouch in which a small amount of barium collected (Fig. 4). Since the time of the operation the patient has done well, although on many occasions, especially when under severe emotional stress, she does develop symptoms very similar to those of which she complained before the operation. Most of those symptoms are controlled by tincture of belladonna after meals. One year later another x-ray study of the lower esophagus demonstrated no significant change (Fig. 5).

*Report by Dr. Henry Brody.

Comment.—It is obvious from the above case study that many difficulties were encountered in finally establishing a definite diagnosis. The original diagnosis of calculous cholecystitis was a deterrent in arriving at our ultimate diagnosis. The patient had symptoms and signs pointing to gall bladder disease for which a cholecystectomy was done in 1948. Following this operation certain symptoms persisted. These might lead one to suspect the diagnosis of a postcholecystectomy syndrome. One symptom, present preoperatively and persisting during the postcholecystectomy course, was the sensation of early bloating following a heavy meal. In spite of the fact that this sensation occurred primarily after emotional stress, it was felt that some organic cause might be responsible. In addition, there was some upper epigastric distress with pain radiating over the substernal area. The diagnosis of coronary artery disease was excluded by a cardiac survey. The ultimate diagnosis rested upon the x-ray examination. This consisted of a series of carefully interpreted x-ray films of the lower esophagus.

Fig. 5.—X-ray picture taken one year later showing no apparent change from Fig. 4.

Surgical intervention was advised as soon as the diagnosis of a tumor in the region of the lower end of the esophagus was made. Because of the anxiety state that existed throughout the ensuing time and because of the patient's marked apprehension concerning any surgical intervention, the operation was delayed until that time when symptoms became progressively worse. At such an opportune moment, surgery was once again recommended and the patient proved amenable to this suggestion. At the time when the patient gave consent to operation she was carefully admonished that, in spite of the fact that operation was indicated to remove a definitely visualized tumor, no guarantee could be given her that all symptoms would be eliminated by such an operation.

At the time of exploration it appeared that we were dealing with a well-circumscribed tumor of the lower esophagus. It is generally accepted that wherever possible these lesions, if they are intramural, should be enucleated. To subject these patients routinely to segmental resection of the esophagus would of necessity expose these patients postoperatively to all the undesirable sequelae of any such resection. Therefore, the lesion was exposed by a longitudinal incision through the outer muscular coat. The lesion was then demonstrated as being intramural, and with finger dissection it was possible to completely enucleate it without entering the esophageal lumen. This was so in spite of the fact that the tumor almost completely involved the entire circumference of the esophagus. After the lesion had been removed it was obvious that there was a marked attenuation of the layers of the esophagus external to the mucosa. It was felt unwise to attempt any sort of plastic repair at this time, since such a procedure might produce marked distortion of the lower end of the esophagus. It was felt that the mucosa would probably accommodate itself to this attenuated muscular layer and in this way obliterate the dead space which was present immediately after operation between the mucosal and extramucosal layers. It was considered unlikely that the muscular layer would shrink down to its original size. Follow-up x-ray films have confirmed our impressions, and while there is obviously a diverticular type of deformity, it in no way produces any obstruction and it empties normally. The fact that there has been no apparent increase in the size of the defect after one year leads us to believe that there will probably not be any increase over the next few years at least. Should the diverticular deformity enlarge and then produce symptoms, it may become necessary to perform some operative procedure on this portion of the esophagus. However, it is our conviction that even if this procedure can be put off for five, ten, or more years the patient is much better off with the deformity that is present than she would be with an esophagogastrostomy. The prognosis for this case, as far as the lesion itself is concerned, is good. Careful microscopic examination of the tumor reveals it to be entirely benign. In view of the fact that the lesion was entirely removed, there is no opportunity for recurrence. On the other hand, symptoms have continued, although to a much milder degree, since the operation. These symptoms are probably more on a functional basis than an organic one. There is no reason for us to believe that these symptoms will change.

DISCUSSION

Benign intramural tumors of the esophagus are usually solitary. Myers and Bradshaw in their review of operated cases found only one in which more than one lesion was present. Most of these tumors occur in the lower third and the majority are leiomyomas. The distribution by sex is almost identical, and the ages of the patients in the literature range from 19 months to 66 years. These lesions usually go unrecognized for many years because of the absence of symptoms. When they become large enough they may produce obstruction or bleeding if the overlying mucosa undergoes necrosis from pressure or

ischemia. In thirty of the forty-two cases reported by Myers and Bradshaw, simple enucleation was done with apparent cure of the lesion. However, in ten cases either esophageal resection or mucosal resection was necessary in order to completely remove the tumor.

The prognosis for these benign lesions of the esophagus, as is true of benign lesions of other parts of the body, is good. When microscopic examination of the lesion after removal reveals that there is no evidence of malignant change, then there is little reason to suspect that there should be any recurrence following total removal. It is well to remember that any one of these tumors while they may appear to be entirely encapsulated and completely benign grossly, may show microscopic evidence of malignancy. The prognosis in those cases is the same as for sarcomas in other portions of the body, the anticipated five-year survival being dependent upon the particular type of sarcoma present. It is not in the province of this paper to philosophize upon the theoretical prognosis of any one of these lesions should they show any malignant change, but it suffices to say that malignant tumors of the esophagus other than carcinomas are extremely rare.

SUMMARY

The case report of a patient in whom an intramural leiomyoma of the esophagus was successfully removed is reported. A brief review of the recent literature is presented.

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GIANT INTRAMURAL LEIOMYOMA OF ESOPHAGUS

A Case Report

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THE case presented here is of interest because it concerns a tumor that is encountered very infrequently, and, in this particular instance the lesion had attained such a great size that its surgical management necessitated an operative procedure of unusual magnitude. This patient was under continuous medical supervision from 1940 until 1952, during which time the esophageal leiomyoma was regarded as a diaphragmatic hernia with a congenitally short esophagus. At the time of surgical resection, a bilateral thoracotomy and an extensive esophagogastrectomy were required to remove the tumor.

CASE REPORT

K. C., Case 315315, was a 17-year-old boy who was admitted to the hospital on May 2, 1952, complaining of dysphagia and the regurgitation of blood. Since the age of 6 he had experienced difficulty in swallowing, and throughout his entire life he had been under constant medical care for this and other complaints. At an early age bilateral congenital cataracts were extracted. He suffered from a congenital hearing impairment and he was required to wear an artificial hearing aid. He had a congenital prolapse of the rectum. Of all difficulties the most annoying to him personally was the inability to eat normally. Because of his multiple handicaps his parents sent him to private boarding schools, and the preparation and ingestion of his food was a ritual that set him apart from his fellow classmates.

The problem of the dysphagia had been studied and treated since the age of 6 both here and at several other larger medical centers throughout the country. X-ray studies (Fig. 1) of the esophagus demonstrated a bizarre picture. The upper esophagus was dilated. The lower half of the esophagus was distorted into a configuration that led to a diagnosis of "congenital shortening of the esophagus with diaphragmatic hernia, stricture, and cardiospasm." These diagnoses were made at more than one medical center and the recommended treatment was always the same, endoscopic dilatation. These procedures were carried out innumerable times from 1940 through 1951 and gave the patient temporary partial relief of the esophageal obstruction. Esophagoscopy had been done on more than one occasion and had shown the esophagus to be narrowed at the mid-esophageal level so that the instrument could not be advanced beyond this point. The mucosa was normal and no tumor was ever visualized.

At the time of this admission diagnostic studies were repeated, and a barium examination of the esophagus was similar to that obtained in 1948. The boy had grown and the structures were larger but the basic picture was unchanged. One border of the tumor can actually be made out just to the left of the midline (Fig. 2). Shortly after this present admission to the hospital the boy's esophagus became completely obstructed. Even water taken by mouth would be regurgitated a few minutes after being swallowed. Repeated attempts were made to introduce a tube into the stomach for alimentation but all were unsuccessful. Efforts were made to pass a string attached to a mercury weighted balloon in order to dilate the area of stricture but this too met with failure as the mercury bag would not drop beyond the mid-esophageal level.

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Operative intervention became mandatory for the maintenance of nutrition, and I elected to make an effort to correct the area of stricture which was believed to be at the point where the thoracic stomach joined the shortened esophagus. On May 24, 1952, a right thoracotomy through the eighth interspace was carried out under endotracheal anesthesia, and upon exploration a large tumor mass was found on the surface of the right diaphragm. It ran obliquely from the lateral chest wall and extended across the mediastinum into the left hemithorax. A tremendously dilated segment of the esophagus entered the surface of the mass from its most superior aspect, and it was soon realized and confirmed by biopsy and frozen section diagnosis that we were dealing with a leiomyoma of the esophagus. The tumor was attached to all contiguous structures with vascular adhesions. These were divided and the mass mobilized as completely as it was possible from

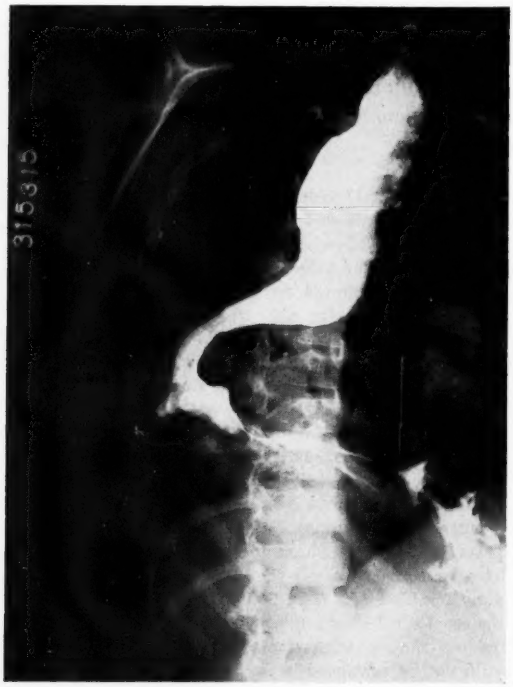

Fig. 1.—Photograph of one of the early esophagrams (Sept. 11, 1948). The tumor mass could not be visualized on the x-ray pictures at this time.

the right-sided approach. The esophagus itself was freed for a considerable distance but subsequently it was found that the dissection of the esophagus should have been carried up much further from the right side. The right thoracotomy incision was then closed, the patient turned, and the operation continued by opening the left hemithorax through the seventh intercostal space. The leiomyoma was then delivered into the left side and the situation evaluated in regard to the extent of resection.

The inferior portion of the tumor constituted its largest diameter and the huge bulbous end extended below the diaphragm. The superior end of the tumor lay just below the aortic arch. The upper end of the esophagus was transected where it entered the tumor, and it was noted in the cut section that the leiomyomatous tissue invaded the wall of the esophagus above the level of the actual tumor mass. It was evident at this time

Fig. 2.—This demonstrates the appearance of the esophagus at the time of the present admission (May 17, 1952). The barium pattern above the right diaphragm was regarded as representing rugal folds of the stomach.

Fig. 3.—Photograph of resected tumor. The upper esophageal end is on the right and the lower gastric end on the left.

that the aortic arch had to be mobilized and the esophagus again divided at a much higher level. Before proceeding with the dissection at the diaphragmatic end, the tumor had to be partially returned to the right chest because it filled the wound and mitigated against any further technical steps. The diaphragm was then incised and the mass found to envelop the upper third of the stomach.

The operation continued and as a preliminary step to resecting the involved portion of stomach, a splenectomy was carried out. The omentum was detached from the greater curvature of the stomach and access gained to the lesser omental cavity. Here, the left gastric artery was isolated and divided between ligatures. The upper end of the gastrohepatic ligament and the attachments of the tumor mass to the diaphragm were cut between clamps and the tumor could then be removed (Fig. 3). The incision in the stomach was closed so as to form an elongated tube that easily reached above the aortic arch without tension. Attention was then directed to mobilizing the aortic arch. Three sets of intercostals and a bronchial artery were divided, and the aorta was retracted anteriorly. The esophagus was delivered above the arch and an additionl 9 cm. of this organ was removed. The cut section at this level showed the muscle layers to be free of tumor tissue, although the wall itself was markedly thickened and edematous. The tissue was of such poor quality that it could be crushed between thumb and forefinger. However, a perfectly tight two layer anastomosis was carried out between the stomach and esophagus. The anastomotic line was placed anterior to the arch and a few inches above it.

The incision in the diaphragm was closed with interrupted sutures and the edge of the new hiatus sutured to the thoracic stomach. A gaping hole was present in the mediastinum, the site from where the tumor had been resected, and a single thoracotomy tube was placed so as to drain both sides of the chest. The left-sided wound was closed and the operation terminated. The total operating time was nine hours. During the course of the procedure the patient was supported with 2,500 c.c. of whole blood and 1,000 c.c. of isotonic saline.

Pathologic examination was carried out in the Department of Laboratories, Henry Ford Hospital, under the direction of Dr. Frank Hartman and was reported, in part, as follows:

"Specimens consists of a large tumor mass, a spleen and portions of stomach and esophagus. The large tumor weighs 1,420 grams and has previously been opened. In the opened condition it measured 19 cm. in length, 15 cm. in width and 7 cm. in thickness."

Microscopically the "tumor is composed entirely of elongated, fusiform, pink staining cells arranged in interlacing longitudinal groups. The nuclei are oval and present very little pleomorphism. The pink-staining cytoplasm is in abundance. This presents a typical picture of leiomyoma."

The patient's life hung in the balance during the first three postoperative days because of extreme respiratory difficulties. He was in an oxygen tent and, in addition, was given supplementary oxygen through a nasal catheter. But even under these conditions he manifested a slight tinge of cyanosis. Any factor that compromised the patient's slender margin of respiratory reserve required prompt attention. A few hundred cubic centimeters of fluid aspirated from one side of the chest or the other would tide him over a period when he was developing respiratory failure. He had considerable bronchial secretion which he was unable to raise efficiently and frequent tracheal aspirations were done to clear the airway. On one occasion a bedside aspiration bronchoscopy was carried out. On the second postoperative day he developed a gastric dilatation which compressed the left lung and resulted in a critical hypoxic state. A gastric tube was passed with prompt alleviation of the dyspnea.

He survived this difficult period and by the end of the first postoperative week he was improving rapidly. Dyspnea was controlled with a small volume of nasal oxygen. A liquid diet was instituted on the fifth postoperative day and was progressively increased with no ill effects. On the eighth postoperative day there was a marked change for the

worse in the patient's condition and it became apparent that he had ruptured the esophagogastric anastomosis and had developed an acute mediastinitis and left empyema. An intercostal tube was inserted into the left chest, and within a few hours 5,000 c.c. of fluid had drained out. Recovery from this point on was relatively uncomplicated.

An attempt was made to treat the esophagopleural cutaneous fistula by feeding the patient through a gastric tube placed in the stomach but this was unsuccessful because a large proportion of the formula would be regurgitated out of the stomach into the fistula.

Fig. 4.—Postoperative esophagram. The upper segment of the esophagus remains dilated.

The method of jejunal alimentation described by Fallis and Barron[1] of this hospital was then instituted. A small intranasal polyethylene tube was passed into the upper portion of the jejunum and a whole liquefied food formula pumped mechanically through the tube into the jejunum. This feeding technique was completely successful in maintaining nutrition and was continued for forty-two days at which time the fistula closed. A small drainage tube was then slowly withdrawn from the residual fistulous tract over a period of one month with eventual complete healing.

At present (nine months later) the patient is completely free of any swallowing difficulties. Postoperative esophagrams (Fig. 4), taken four months after operation, show a satisfactory result. The upper esophagus is still dilated but there is no delay in the passage of barium through the anastomosis into the thoracic stomach.

DISCUSSION

In recent years several authors have pointed out the rarity with which benign tumors of the esophagus are encountered. In an article published in

1950 Chi and Adams[2] tabulated the benign esophageal tumors that had been treated surgically and reported in the medical literature. These were of all types and totaled only twenty at that time. Their article also contains an excellent bibliography on this subject.

In the majority of the cases cited, the lesions were of the mucosal pedunculated type or else they were localized intramural myomas that could be enucleated from the esophageal wall without interrupting the continuity of the organ. There are only three instances recorded in the medical literature where

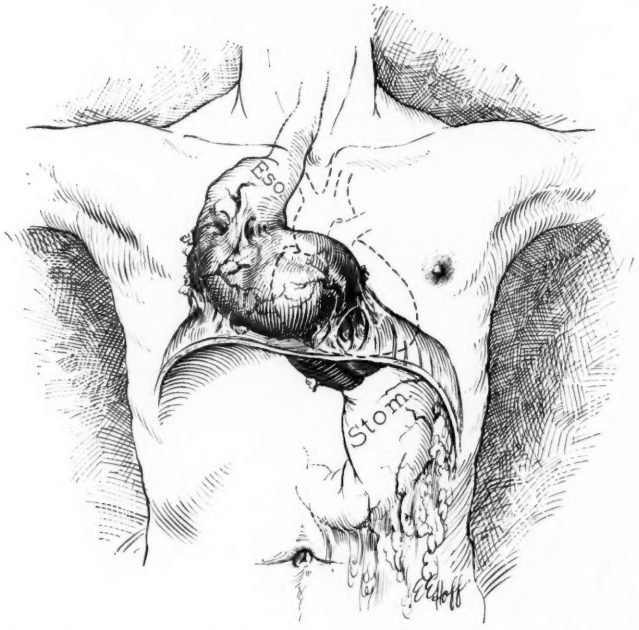

Fig. 5.—Artist's diagrammatic reconstruction of the esophageal leiomyoma in situ illustrating the relations of the tumor.

intramural leiomyomas of the esophagus were successfully treated by a radical esophagectomy and reparative esophagogastrostomy. Broch[3] (1942), Kenworthy and Welch[4] (1948), and Chi and Adams[2] (1950) described cases that required an extensive surgical resection. In Broch and Kenworthy's cases the leiomyomas involved the esophagogastric junction. The patient operated upon by Chi and Adams had a leiomyoma in the upper third of the esophagus. In the case described here, the tumor involved the lower two-thirds of the esophagus proper, including the cardia and the upper third of the stomach.

A correct preoperative diagnosis was not made in this case and the decisions regarding management had to be made at the operating table. Two problems were immediately considered after the true nature of the pathology was ascertained and before the mobilization of the tumor was begun.

The first problem was the necessity of carrying out a bilateral thoracotomy in one consecutive operative procedure. Study of Figs. 5 and 6 will illustrate why exposure from both right and left sides of the chest was mandatory in the technical management of this case. The bulk of the tumor mass was situated in the right hemithorax, but its largest end was on the left side and it descended into the left upper quadrant of the abdomen. A supplementary abdominal incision would not have provided adequate exposure. Consequently we were forced to open the left side and incise the diaphragm for proper visualization in this area. Chi and Adams[2] opened both sides of the chest in their patient, but the two thoracotomies were separated by a month's interval.

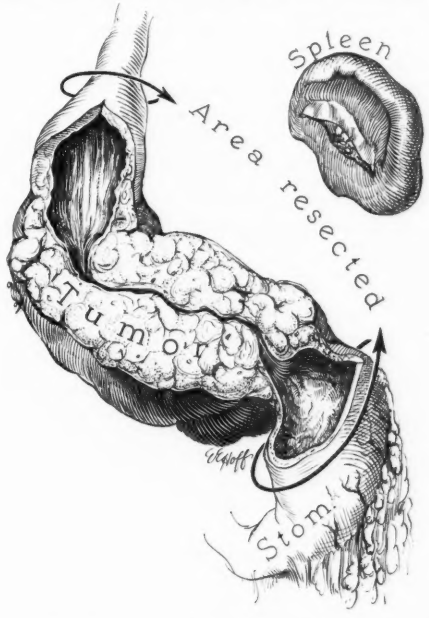

Fig. 6.—Cross-sectional drawing showing the extent of surgical resection.

I was aware of Overholt's[5] experience, where, in three instances, simultaneous bilateral thoracotomies had been done. Encouraged by these previous successes, the decision was made to proceed with the operation from this standpoint.

Overholt's patients experienced no serious problem in their postoperative physiologic adjustment to simultaneous bilateral thoracotomy. In the case presented, the physiologic respiratory insult was extremely marked. The patient survived only because the stamina of youth was in his favor and because of the diligent care given by the surgical residents. The duration of the operation, the fact that the thoracotomies were consecutive and not simultaneous, and the extensive surgical resection that was required undoubtedly accounts for the severe postoperative reaction in this patient.

The second problem considered at the operating table was a definite deterrent to proceeding with the removal of the tumor. Because the esophagus above the mass was dilated and its wall was markedly edematous as a result of the chronic obstruction it was questionable whether a leak-proof gastroesophageal anastomosis could be made. Even if this were accomplished it seemed doubtful whether or not tissue of this nature would heal. The opinion of Dr. Laurence S. Fallis, Surgeon-in-Chief at this hospital, was sought in this regard. He felt that there was no other acceptable alternative in the management of the case, except resection, and advised that the risk of carrying out the anastomosis should be assumed.

The anastomotic breakdown which occurred on the eighth postoperative day was not unexpected, and was diagnosed a few hours after it happened. Prompt drainage was instituted, massive doses of antibiotics started, and the patient recovered rapidly from the acute effects of the accident. Subsequent course was remarkable for the manner in which nutrition was maintained in the face of an esophageal fistula and a draining empyema. Throughout the forty-two day period, when nothing was administered by mouth, the patient lost only five pounds in weight. Dr. James Barron, of this hospital, managed this aspect of the case and he estimates that between 4,000 and 5,000 calories a day were pumped through the polyethylene tube into the jejunum.

SUMMARY

A case of esophageal leiomyoma is reported. This is believed to be the fourth such case recorded that survived a radical esophagectomy and esophagogastrostomy. It is thought to be the largest esophageal tumor (1,420 Gm.) ever resected surgically.

The method of jejunal alimentation which was used in the management of the esophagopleural-cutaneous fistula that developed in this patient was eminently successful in maintaining nutrition.

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INTRATHORACIC MENINGOCELE NOT ASSOCIATED WITH NEUROFIBROMATOSIS

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INTRATHORACIC meningoceles are of sufficient rarity to warant the presentation of individual cases. Only fourteen proved cases could be found in the literature at this time. The following case, we believe, is the fifteenth proved case and the fourth case not associated with neurofibromatosis.

CASE REPORT

A 29-year-old Negro woman (gravida iii, para i) was seen in the emergency room on April 4, 1952; she complained of dysmenorrhea and menorrhagia. A chest film revealed a large intrathoracic mass, apparently unassociated with these symptoms. She was referred to chest clinic but failed to keep the the appointment. Two months later she was readmitted to the hospital with the same essential complaints, was treated, and then transferred to the chest service.

Past history and family history were noncontributory. Physical examination was essentially negative except for the presence of bilateral lower quadrant tenderness and a large midline mass extending almost to the umbilicus. A mass in the right uterine adnexa deformed the vaginal vault. These masses, surgically removed at a later date, were fibromyomas of the uterus. No evidence of neurofibromatosis was noted. The laboratory findings, including spinal fluid studies, were normal.

Chest films revealed a large, sharply circumscribed, homogeneous density filling the entire upper third of the left lung field (Fig. 1). The trachea was displaced slightly anteriorly, and the esophagus was displaced to the right. A scoliosis of the upper thoracic spine was present, with the concavity on the left side. No definite bone destruction was noted but there was thought to be pressure atrophy of the left first rib posteriorly. A lung cyst or neurofibroma was considered the most likely diagnosis by both the surgeons and radiologists.

Operation.—A thoracotomy was done on the left side through the bed of the fourth rib. A round, cystic tumor, approximately 10 cm. in diameter, was seen to fill the apex of the left chest. The pleura and the second intercostal nerve on the anterior surface of the tumor were dissected away, and approximately 150 c.c. of clear fluid was aspirated to facilitate removal of the tumor. Dissection was carried down to the second intervertebral foramen, through which the mass entered the chest, and the cyst was opened. The foramen was only slightly larger than normal, measuring approximately 7 or 8 mm. in diameter. Through the lumen of the cyst pedicle could be seen the dentate ligaments and spinal cord. At this point, it was evident that the cyst was actually a meningocele. The neck of the meningocele was closed with a purse string suture of 000 silk reinforced by interrupted sutures of No. 30 cotton. The suture line was covered with Gelfoam.

Postoperatively the patient drained a light amber fluid, thought to be spinal fluid, from the wound at the rate of 300 to 350 c.c. daily. By the third week this decreased to 125 c.c. daily, gradually diminishing thereafter until it finally ceased. The pleural drainage tubes were then removed with no reaccumulation of fluid. There was a slight weakness of the left arm and left leg which cleared within a few days. This was thought to have been due to arachnoiditis. A definite Horner syndrome was noted on the left. This was said to have been present to a lesser degree preoperatively though not recorded on the

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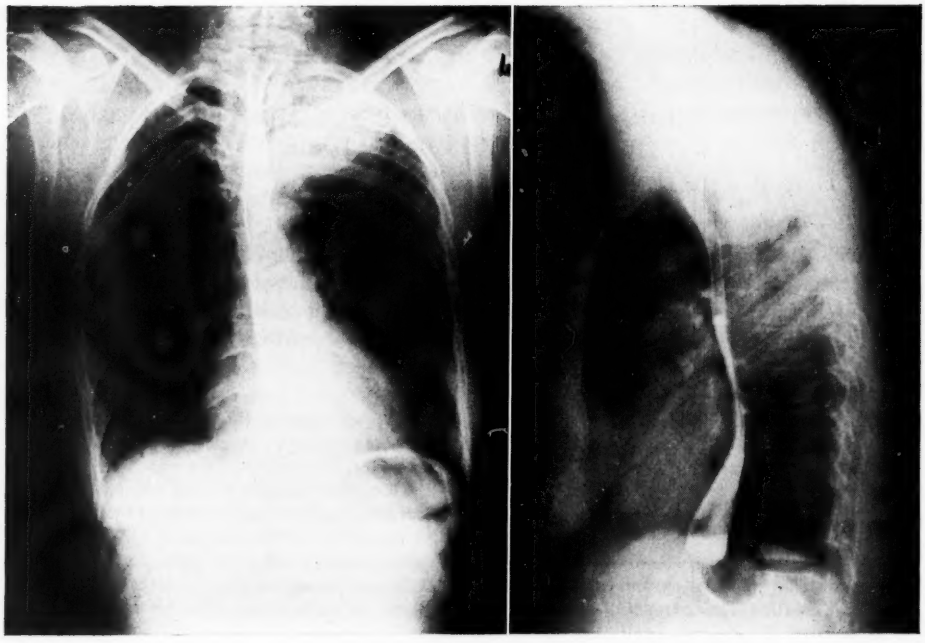
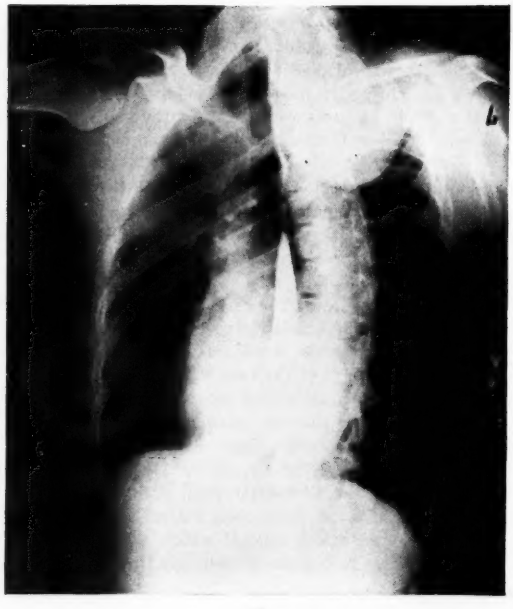


A. *B.*



C.

Fig. 1.—Radiographs taken in the posteroanterior (*A*), left lateral (*B*), and left anterior oblique (*C*) positions show a mass in the apex of the left lung field.

chart. The patient was given penicillin and streptomycin prophylactically. Upon discontinuance of these drugs the patient's temperature rose to 102° F. and she developed a stiff neck, headache, and return of the weakness of the left leg. A lumbar puncture at this time revealed normal findings. Upon reinstitution of antibiotic therapy, the fever promptly abated and the meningeal signs and symptoms disappeared. The weakness of the left leg persisted, however. Two and one-half months following surgery the patient was able to walk without the aid of a cane but still exhibited a slight bilateral ankle clonus, hyperactive ankle jerk, and weakness of the hamstring group of muscles of the left leg.

A summary of all proved cases reported to date with an additional two cases not previously tabulated is presented below.

DISCUSSION

In reviewing the cases summarized in Table I the constancy of certain clinical features was striking. Ten of the fifteen cases had neurofibromatosis; all but one had definite vertebral abnormalities. These two findings in the presence of an intrathoracic mass, paucity of symptoms, neurologic or otherwise, should stimulate efforts to demonstrate a meningocele by myelography. In two of these cases the correct diagnosis was made prior to surgery, 2 and 6. The most frequent preoperative diagnosis was that of neurofibroma.

TABLE I. SUMMARY OF CASES PREVIOUSLY REPORTED*

AUTHOR	NO. OF CASES	TREATMENT	ASSOCIATED NEUROFIBROMAS	VERTEBRAL CHANGES
1. Pohl,[1] 1933	1	Surgery; death from empyema	Yes	Yes
2. Schüller,[2] 1938	1	Not resected; death following neurectomy	Yes	Yes
3. Ameuille,[3] 1940	1	Surgery; cyst packed; death from empyema	Yes	Yes
4. Welch,[4] 1949	1	Resection; good results	Yes	Yes
5. Byron,[5] 1949	3	One case resected with good results. One case treated conservatively. One case resected; the patient expired during surgery	Yes, two cases No, one case	Yes
6. Mendelsohn[5]	1	Resection; good results	Yes	Yes
7. Head[5]	1	Treated conservatively	?	?
8. Ciaglia[5]	1	Resection; good results	Yes	Yes
9. Cross,[6] 1949	1	Treated conservatively	No	Yes
10. Kessel,[7] 1951	2	One case resected with good results. One case treated conservatively	Yes, both	Yes
11. Rubin,[8] 1952	1	An infant who expired before treatment was instituted.	No	Yes

*Campos[9] in 1946 presented two cases with the above findings but the diagnosis was not definitely proved. In view of the frequency of the associated findings, they probably represented meningoceles.

The importance of the associated clinical features was pointed out by Rubin[8] in a review of two separate series of intrathoracic neurogenic tumors. Only four cases of neurofibromatosis were found in a total of 135 cases. This is an important observation because in almost all of the cases presented here, the provisional diagnosis was that of neurofibroma.

No etiologic factor responsible for the defect has been definitely established. Most authors agree that a developmental defect of the bone or dura, or both, precedes the appearance of the meningocele.[6] Which of these defects is more important is likewise undetermined.

Treatment by excision seems to be fairly safe with modern surgical techniques. Resection was not attempted in two cases because of lack of symptoms and the relatively safe location of the tumor.[5] The choice of treatment must of necessity be individualized.

SUMMARY

1. A case of intrathoracic meningocele is reported; we believe it to be the fifteenth proved case in the literature.

2. Roentgen findings common to most of the cases of this type are kyphoscoliosis, a mediastinal tumor, evidence of erosion of either the vertebral bodies or adjacent ribs, and vertebral deformities or anomalies. An important associated clinical finding is the presence of neurofibromatosis which was present in ten of the fifteen proved cases comprising this series. Neurofibromas associated with a tumor located paraspinally in the chest should initiate a search for meningocele by myelography.

ADDENDUM

Since this article was written, another case of intrathoracic meningocele proved by aspiration has been recorded. Ottani, Gerardo: Intrathoracic Meningocele, Ann. di radiol. diag. **23**: 416-419, 1951. (Abstracted in Radiology **59**: 132, 1952.)

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RESULTS OF SUPERVOLTAGE ROENTGENOTHERAPY OF ESOPHAGEAL CARCINOMA

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WE HAVE repeatedly discussed the rationale of supervoltage roentgenotherapy for epidermoid carcinoma of the esophagus, its indications, contraindications, and technical details. More recently we have presented a comparison of the optimal surgical and radiologic results obtained to date.[5] The following article is a summary of our own results in patients treated between January, 1940, and July, 1952. For technical details of treatment we refer to our previous publications.[1, 3, 4]

A total of fifty-eight patients was seen (Table I). Nineteen of these were not treated, in most instances because either the lesion was too advanced to expect even temporary sustained palliation, or the general condition was too poor to support such a major procedure. Twelve patients received what we would consider grossly inadequate therapy, usually because treatment had to be discontinued on account of the rapidly declining general condition.

Twenty-seven patients received what we would consider adequate treatment. By this we mean that a minimal dose of 5,000 r., calculated at the level of the esophagus, in about forty to forty-five days, or its equivalent with greater fractionation, was delivered. Of these patients, five are in clinically satisfactory condition today.

Considering only patients who have been treated more than five years ago, twenty-nine were seen of whom five received incomplete, and fourteen complete treatment, as previously defined. Three patients are well without radiographic or clinical evidence of disease. They have been living a perfectly normal life in every respect for periods of twelve, ten, and eight years, respectively. One of these had disease localized in the cervical segment, one in the upper, and one in the lower thoracic segment of the esophagus. Thus three patients have been well for more than five years out of a total of twenty-nine patients examined, or out of nineteen patients who were admitted for treatment (corresponding to surgical exploration), or out of fourteen who were completely treated (corresponding to patients in whom surgical resection would have been completed). In addition, two patients have been clinically well, possibly with controlled disease for two and three years, respectively. One had a carcinoma in the upper thoracic and one in the mid-thoracic portion. These two patients do not have restoration of a normal esophagus insofar as both have a moderate degree of obstruction. This, however, has remained entirely stationary in one patient for fifteen months, in the other for eighteen months. Since the stenoses in these patients have not

From the Tumor Institute of the Swedish Hospital.
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TABLE I. EPIDERMOID CARCINOMA OF ESOPHAGUS

	NO. SEEN	NOT TREATED	INCOMPLETELY TREATED	COMPLETELY TREATED	WELL JULY 1, 1952
January, 1940—July, 1952	58	19	12	27*	5†
January, 1940—July, 1947	29	10	5	14	3‡

*Histologically verified by biopsy or autopsy.
†Two patients well two and three years, respectively.
‡One lesion in cervical, upper thoracic, and lower thoracic segment, respectively.

changed over this long period of time and have remained radiographically smooth, we believe that they are more likely caused by fibrotic scarring than by active disease. One of these patients needs occasional dilatation. Both feed themselves satisfactorily by semisolids.

None of the patients with lesions in the mid-thoracic portion of the esophagus has remained well. The question then arises whether surgery would have offered any of these patients a better chance of control.

An analysis of the twenty-two failures shows that in eight instances roentgenotherapy accomplished re-establishment of the normal esophageal passage to the point that these patients could eat normally, or almost normally, up to the time, or very shortly prior to the time, of death. Death was not due to recurrent obstruction or demonstrable recurrent disease in the site of primary origin. The analysis of the cause of death in these eight patients (Table II) shows that in five instances failure of permanent control was due to the presence and progression of paraesophageal and mediastinal nodes; in one patient death was due to liver metastases. In one patient, death thirteen

TABLE II. EPIDERMOID CARCINOMA OF ESOPHAGUS. CAUSE OF DEATH IN PATIENTS IN WHOM FOLLOWING IRRADIATION NORMAL OR ALMOST NORMAL ESOPHAGEAL PASSAGE WAS MAINTAINED UNTIL DEATH OR UNTIL VERY SHORTLY PRIOR TO DEATH

INITIALS	NO.	AGE	LOCATION	DURATION SYMPTOMS PRIOR TREATMENT (MO.)	DOSE* R./DAYS	DURATION END OF TREATMENT TO DEATH (MO.)	CAUSE OF DEATH
J. R.	41-	62	M	12	(16200/81)	25	Fibrosis
G. G.	44-	58	L	14	(14000/61)	15	Paraesoph. node 1 yr. p. rad.
B. S.	46-740	50	C	4	6500/52	6	Paraesoph. node
I. G.	47-456	61	L	16	5000/44	19	Liver metastasis
D. B.	48-341	76	C & U	7	(7100/30)	9	Extrins. obstruct. node lower esoph. 9 mo. p. rad.
M. P.	50-22	55	C	2	6000/36	6	Mediast. nodes
J. O.	50-620	76	M	3 wk.	6000/39	13	General weakness; age
O. W.	51-975	60	M	2	7000/45	5	Perfor. of mediast. node; acute massive hemorrh.

C—Cervical segment.
U—Upper thoracic segment.
M—Mid-thoracic segment.
L—Lower thoracic segment.

*Dose in parentheses is measured at skin in patients in whom chest measurements are not available for computation of tumor dose. Figures of dose without parentheses represent tumor dose as calculated at level of esophagus.

months after completion of treatment without clinical or radiographic signs of active disease in esophagus, mediastinum, or elsewhere was attributed to general weakness and age. Obviously, surgery would not have had any better chance of success in any of these patients. In only one instance can it be argued in retrospect that surgery might have been more successful. This patient died twenty-five months after very massive therapy from what we believed was probably massive mediastinal and pulmonary fibrosis rather than active disease. This patient was treated in 1941, a time when resection of the mid-esophageal portion still carried a prohibitive surgical risk. Even in this instance, it remains doubtful what surgery could have accomplished in the presence of a disease which had caused symptoms necessitating regular dilatation for more than one year prior to referral to us.

Thus it appears that in these eight patients, roentgenotherapy has accomplished satisfactory palliation preventing starvation for periods of five to twenty-five months during which time these patients lived practically normal lives, in one-half of the instances for periods exceeding one year.

The analysis of the remaining fourteen failures shows that in these instances the primary disease remained uncontrolled and progressed. Perforation into the tracheobronchial tree, mediastinum, or aorta one to seven months following completion of therapy was the cause of death in six patients. In two cases death was due to generalization of the disease, and in one instance to ascending urinary infection secondary to a cord bladder caused by vertebral involvement by metastatic disease one month after completion of therapy.

In these fourteen cases in which the primary disease progressed following radiation therapy, the question of whether surgery could have accomplished more becomes even more urgent. Six of these patients were referred to us by three of the leading thoracic surgeons in the community because they were considered inoperable: four because of demonstrable extraesophageal extension (radiographically demonstrable penetration or mediastinal nodes); one with an anaplastic carcinoma of 10 cm. length with the history of symptoms for more than nine months; one because of the general condition and age. Three additional patients were considered inoperable because of general condition and age, one of these with fibrillation prior to therapy, and one with Parkinson's disease. Two patients had extensive cervical and mediastinal lymph node involvement prior to therapy. One patient had a carcinoma extending from the inferior level of thyroid cartilage to the fourth dorsal interspace. The patient previously mentioned, who developed metastatic involvement of one vertebra with cord bladder one month after treatment, would obviously not have been benefited by surgery. There remains one patient, aged 72 years, with a lesion at the level of the sixth thoracic vertebra in whom a very aggressive thoracic surgeon may perhaps have considered surgery (C. J., 44-). The last patient to be accounted for (J. P., 47-477) was 58 years old and in good general condition at the time of admission; he was offered the choice of surgery with its inherent risk and an attempt with radiation therapy. Two months following the completion of treatment, this lesion appeared uncontrolled although it could not be determined whether the findings were due

to active disease or to massive radiation reaction and necrosis. At that time, therefore, it was felt that the likelihood of uncontrolled disease was sufficiently great to justify the risk of surgery. Resection of a 9 cm. mid-esophageal segment with primary esophagogastrostomy was done. In spite of the heavy preceding irradiation (4,800 r. in forty days calculated at the esophageal level) healing progressed without complications and the patient was discharged ten days following surgery. No carcinoma was found in serial sections of the removed specimen. Six days after discharge from the hospital, the patient was killed in an automobile accident. At autopsy carcinoma was found infiltrating the esophageal wall about 5 mm. above the site of the anastomosis and a carcinomatous nodule 1 cm. in diameter at the level of the anastomosis.

Thus it would seem that none of the patients not saved by radiation therapy would have had a better chance if surgery had been elected.

SUMMARY

1. Between January, 1940, and July, 1952, fifty-eight patients with carcinoma of the esophagus were seen. Five out of twenty-seven adequately treated, or of thirty-nine patients in whom treatment was started, are clinically well today.

2. Of twenty-nine patients seen more than five years ago, nineteen were admitted for treatment and fourteen of these received adequate therapy. Three of these patients have been clinically and radiographically well, without any sign of active disease with normal esophageal function, for periods of twelve, ten, and eight years, respectively.

3. Of twenty-two patients adequately treated but uncontrolled, normal or almost normal esophageal function until death, or until shortly prior to death, was accomplished in eight instances for periods between five and twenty-five months. In six of these patients death was due to extraesophageal progression of the disease, in one to general weakness caused by age, and in one probably to massive mediastinal and pulmonary fibrosis.

4. Analysis of the twenty-two adequately treated but uncontrolled cases suggests that in only one of them surgery may possibly have offered a better chance.

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Announcements

THE AMERICAN ASSOCIATION FOR THORACIC SURGERY

Notice of Annual Meeting

The 34th annual meeting of the American Association for Thoracic Surgery will be held May 3, 4, and 5, 1954, in Montreal, Que., Canada. Headquarters will be at the Sheraton-Mount Royal Hotel.

Requests for Hotel Reservations

These should be mailed *directly* to the Reservation Manager, Sheraton-Mount Royal Hotel, Montreal, Que., Canada. Please mention this Association, type of accommodation desired, date, and approximate hour of arrival. If accommodations are desired elsewhere, please communicate directly with hotel of your choice.

Thoracic Surgical Forum

After due consideration, the Council has decided to try an experiment by assigning one morning session to a "Thoracic Surgical Forum" for the presentation of short papers on current thoracic research, anatomy, modified surgical techniques, etc. Papers at this session will be limited sharply to ten minutes (including the showing of lantern slides or other illustrative material) with three minutes as the maximum discussion of each paper.

These papers will be published together in one issue of THE JOURNAL OF THORACIC SURGERY. Maximum length: 3,000 words without illustrations. If illustrated, an appropriate number of words must be subtracted for each cut used.

Abstracts for Papers

Abstracts for the presentation of papers at the 1954 meeting, including papers before the Forum, must be received on or before January 1, 1954, otherwise they will not be considered by the Program Committee.

Abstracts should be labeled "For Forum" or "For Regular Program."

Five, repeat five, copies of each abstract should be sent to the Secretary of the Association, Dr. Paul C. Samson, 2938 McClure St., Oakland 9, Calif. They should not be sent to the Editor.

The abstracts should contain from 200 to 250 words and accurately reflect the contents of the completed paper.

Applications for Membership

Applications for Associate Membership in the Association must be received by the Membership Committee not later than Jan. 1, 1954, to be considered for action in Montreal. Applications received beyond this deadline will be deferred for consideration until 1955.

Applicants must be sponsored by three Active or Senior Members of the Association. In addition to the sponsoring statement on the application form, the sponsors *must* submit a separate letter concerning the applicant direct to Dr. Lyman H. Brewer, III, Chairman of the Membership Committee, 2010 Wilshire Blvd., Los Angeles, Calif. Completed application forms should likewise be sent to Dr. Brewer. These regulations are at the direction of the Council and were made effective in 1952 at Dallas.

Greetings to Thoraxchirurgie

The JOURNAL of THORACIC SURGERY takes pleasure in welcoming to the same field *Thoraxchirurgie* of which the first number has appeared under date of April, 1953. The new journal will be devoted to clinical and operative surgery, pathologic physiology, experimental pathology of the thoracic viscera, and to anesthesia.

It will be under the editorial direction of Professor K. Vossschulte of Giessen, assisted by A. Lezius of Hamburg and R. Nissen of Basel. The all-European editorial board contains the names of several thoracic surgeons well known to Americans, among whom are Brock, Crafoord, Denk, Dogliotti, Felix, and Santy. *Thoraxchirurgie* will be published by the experienced firm of Georg Thieme of Stuttgart, the publishers of *Deutsche medizinische Wochenschrift*, and an issue will appear every two or three months.

The first number, with a foreword by Nissen, makes a splendid impression. It is gratifying that a new journal has appeared devoted to thoracic surgery which will be published in the language used by Sauerbruch for his pioneer contributions to this field.

The Editor